

Non-inflammatory pathologies of the middle ear

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Content

- Otosclerosis
 - Case presentation
 - State of the art
- Differential diagnoses in cases with conductive or mixed hearing losses
- Discussion

Third Edition

Schuknecht's
PATHOLOGY
of the
EAR

Henning Hildmann
Holger Sudhoff

Middle Ear Surgery



DVD-VIDEO



INCLUDED

 Springer



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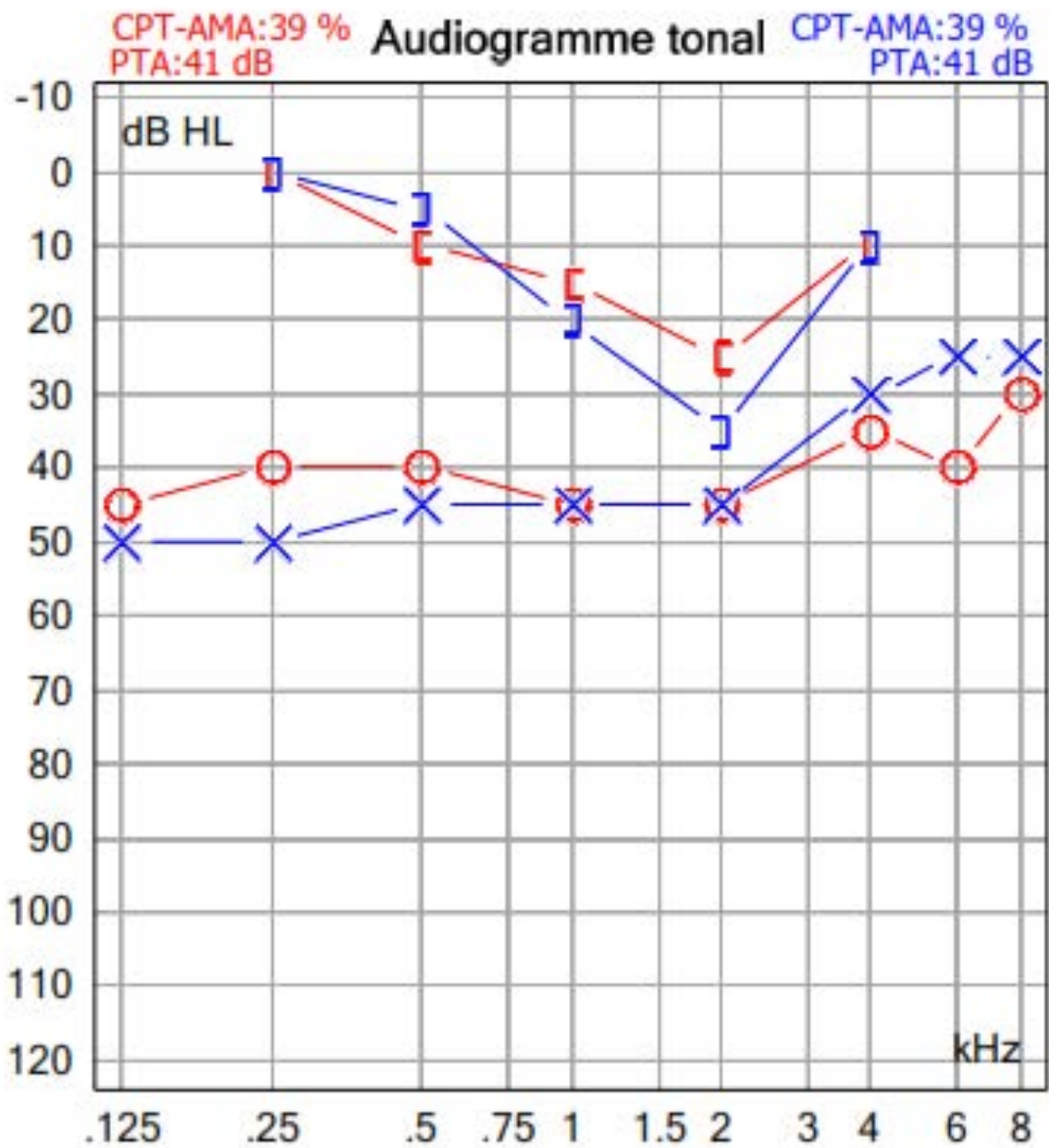


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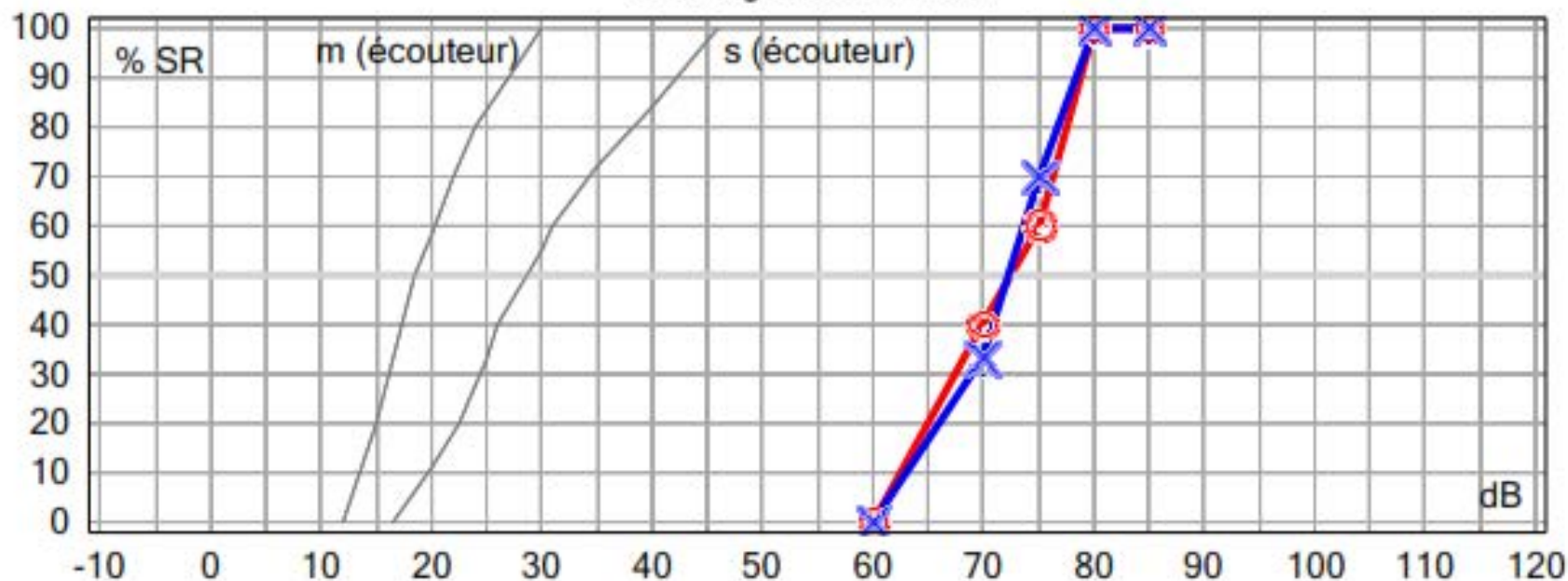
FACULTÉ DE MÉDECINE

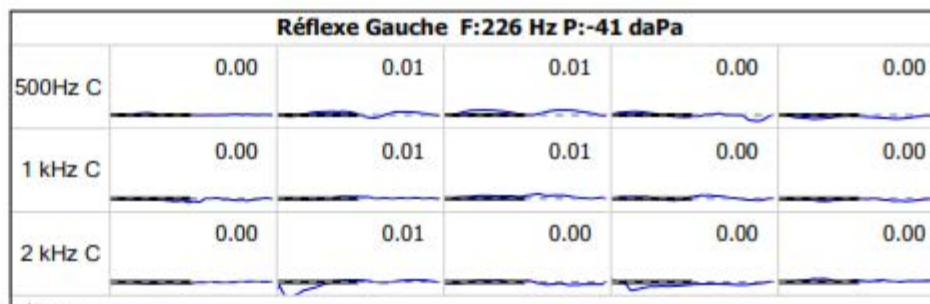
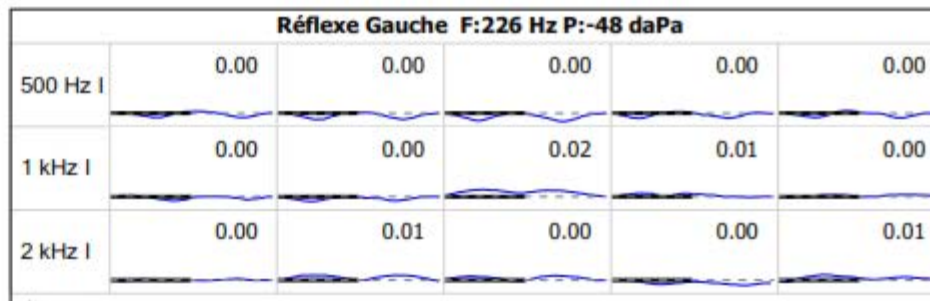
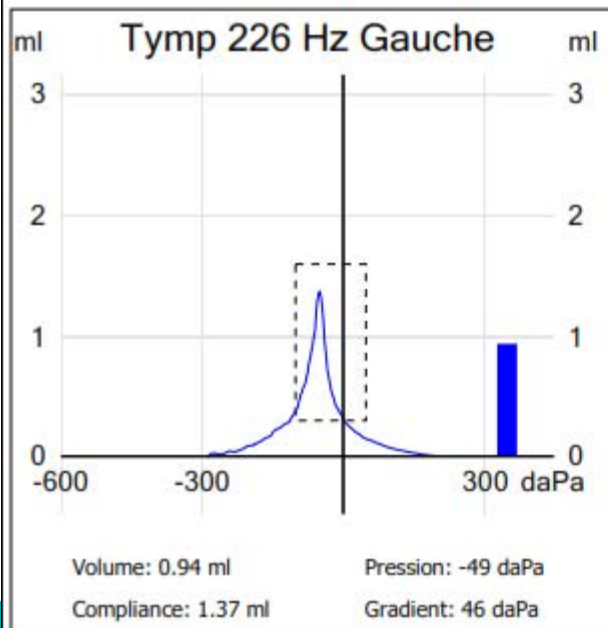
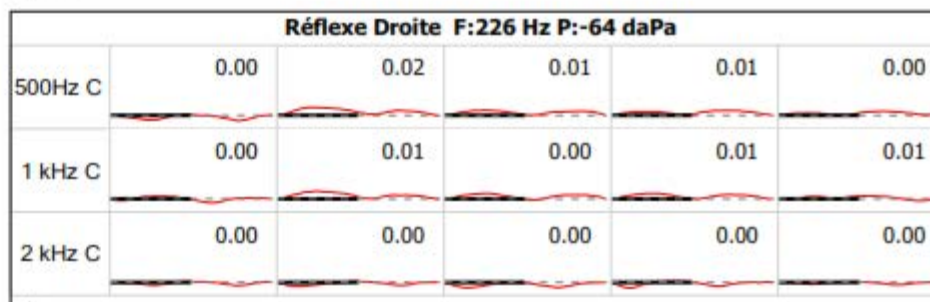
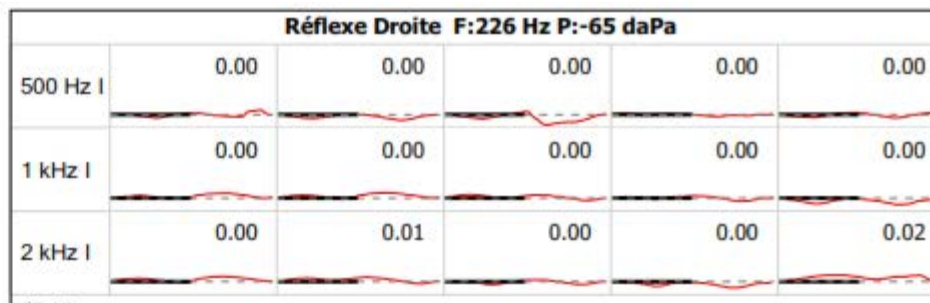
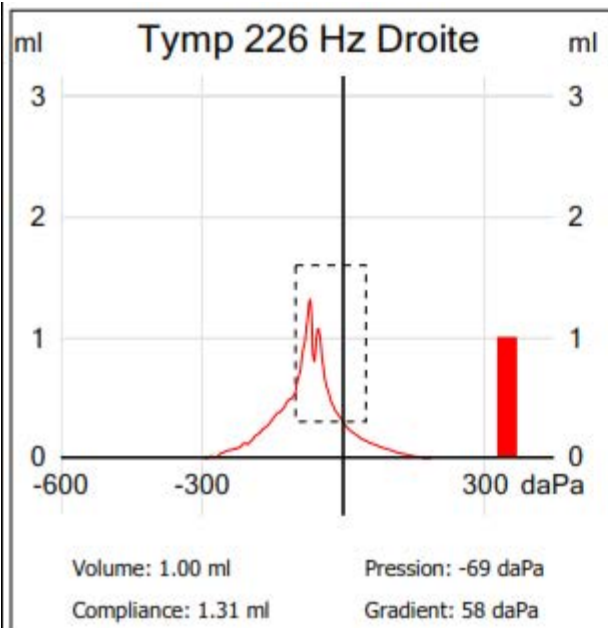
39-year old bank employee

- Bilateral hearing loss since about 3 – 4 years without other symptoms
- Has bought a hearing aid for the left side 2 years ago, but is not satisfied
- Mother and sister also hearing impaired, have been operated successfully in the past
- Status: Weber centered, Rinne négatif bilatéralement, tympaniques normales



Audiogramme vocal





Do we need a CT-scan preoperatively?

Arguments against CT

- Radiation exposure¹
 - 1 CT scan triples the risk for leukemia and brain cancer in children
 - 1 excess cancer in 10'000 CTs at childhood age
- No additional clinical information²
- Costs

¹Pearce M et al. Radiation exposure from CT scans in childhood and subsequent risk of leukaemia and brain tumours: a retrospective cohort study. Lancet. 2012 Aug 4;380(9840): 499-505. (cohort size of 180'000 people)

² Wegner L et al. Otol Neurotol. 2016 Jan;37(1):9-15

A Systematic Review of the Diagnostic Value of CT Imaging in Diagnosing Otosclerosis.

Arguments for CT

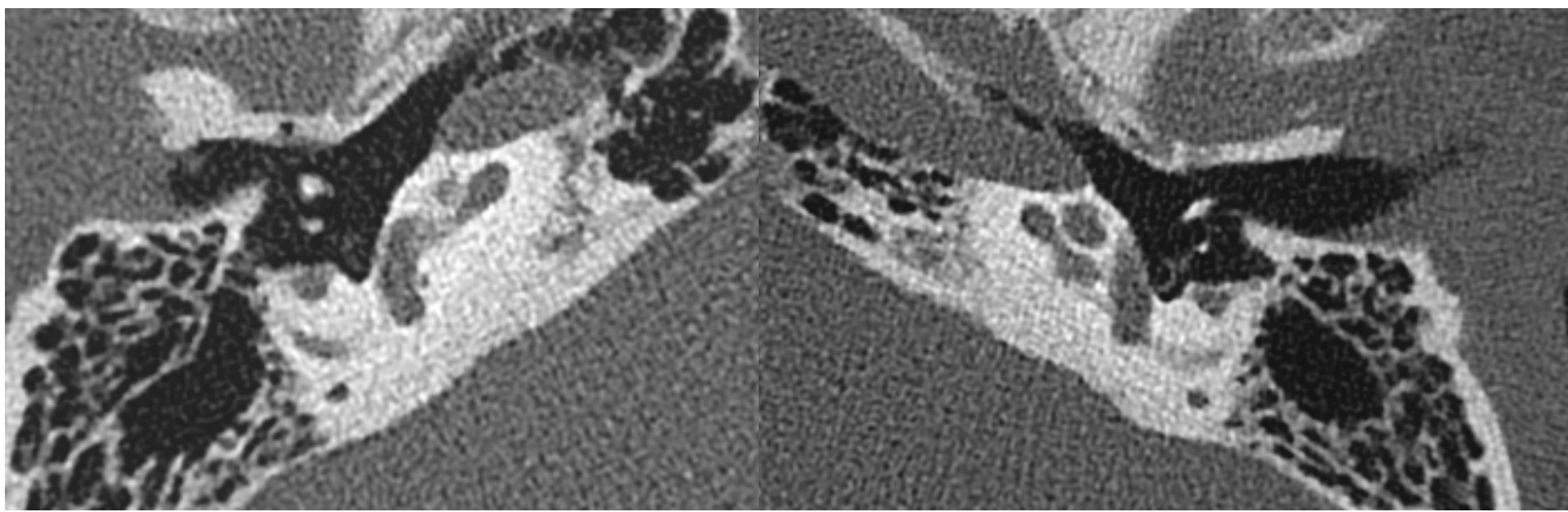
- Confirmation of diagnosis¹
- Ruling out of differential diagnoses¹
- Anticipating potential complications
 - Floating footplate¹
 - Gusher ear²
 - Anatomical (facial nerve) abnormalities

¹Lagleyre S et al. Otol Neurotol. 2009 Dec;30(8):1152-9. Reliability of high-resolution CT scan in diagnosis of otosclerosis.

²McFadden et al. Ear Nose Throat J. 2005 Dec;84(12):770, 772-4.

Preoperative computed tomography may fail to detect patients at risk for perilymph gusher.

Where is the otosclerotic lesion?



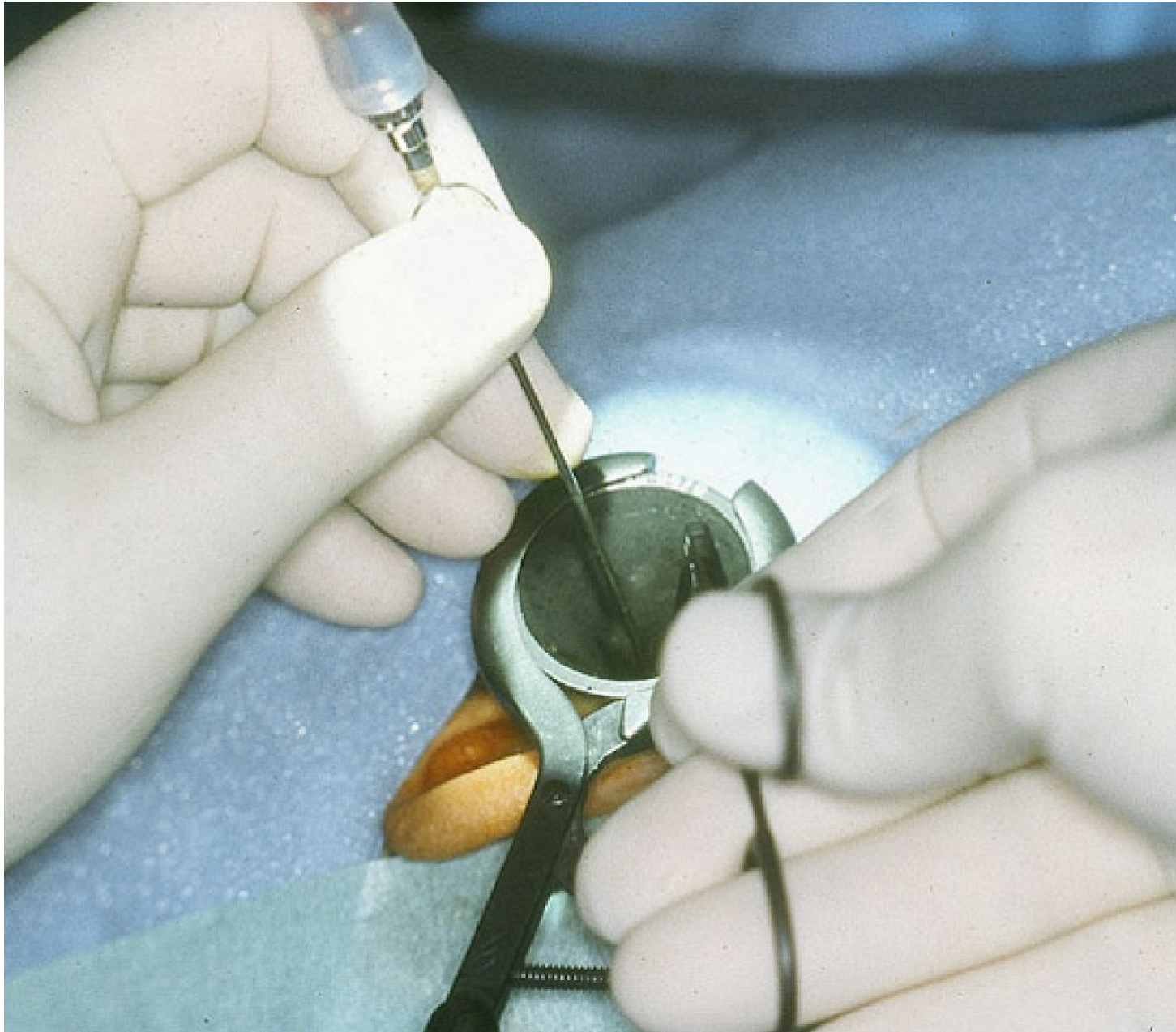
39-year old bank employee

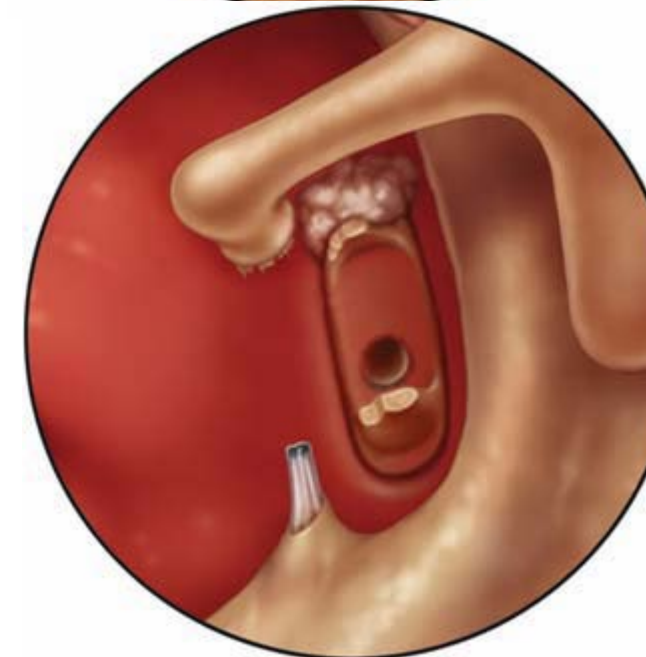
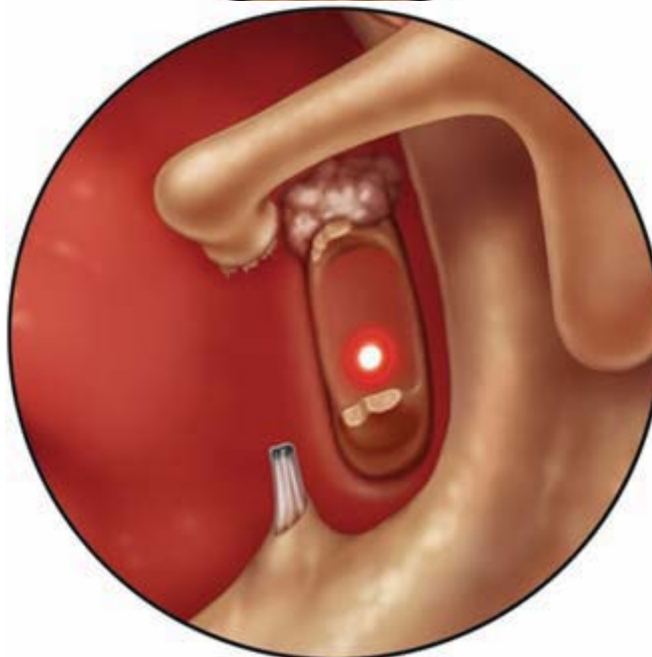
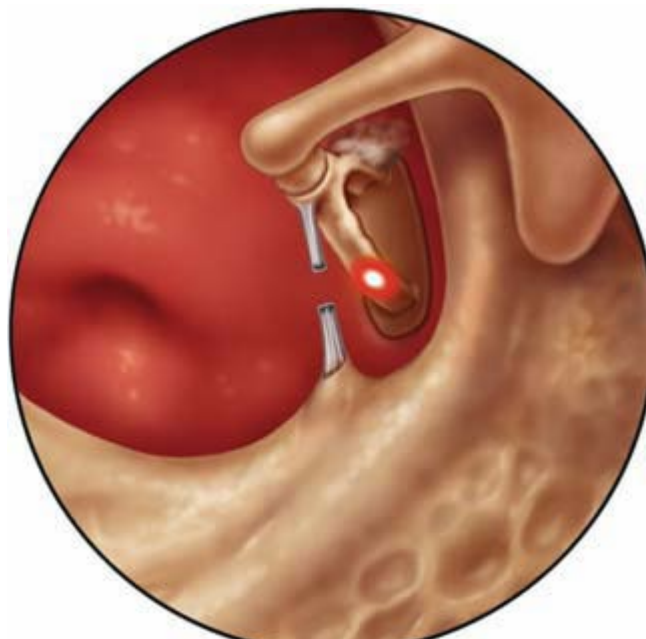
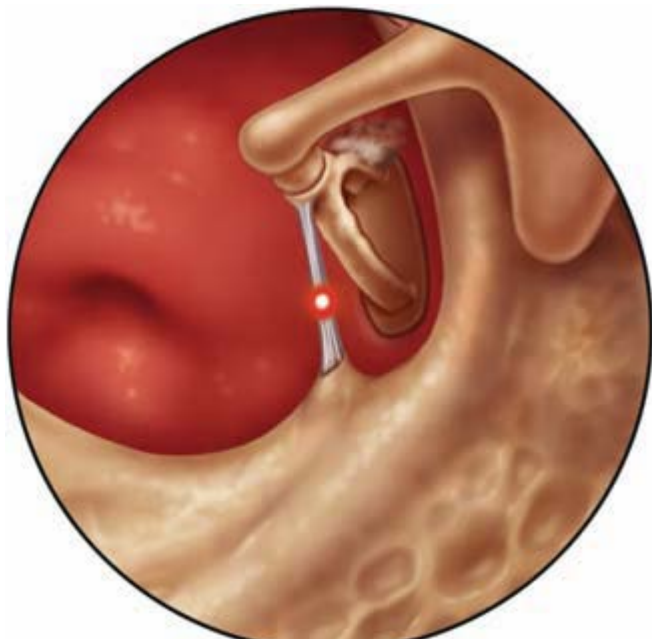
- Conductive hearing loss without stapedial reflex and positive familial history of otosclerosis

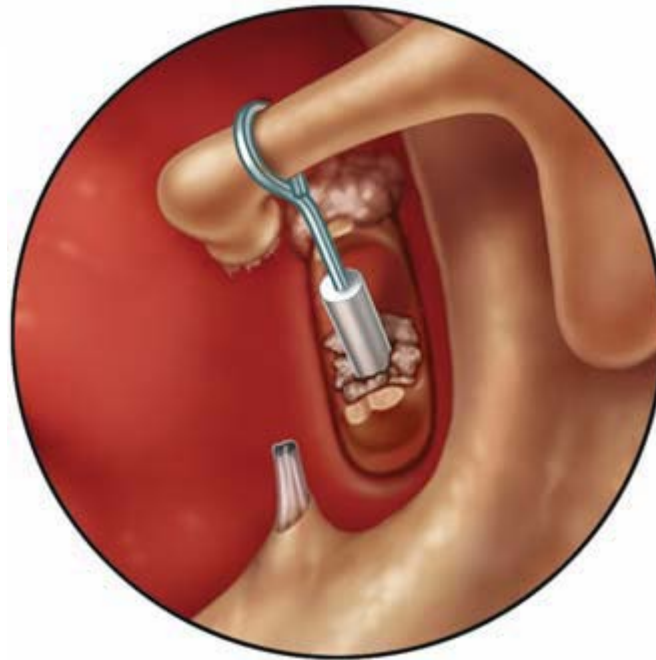
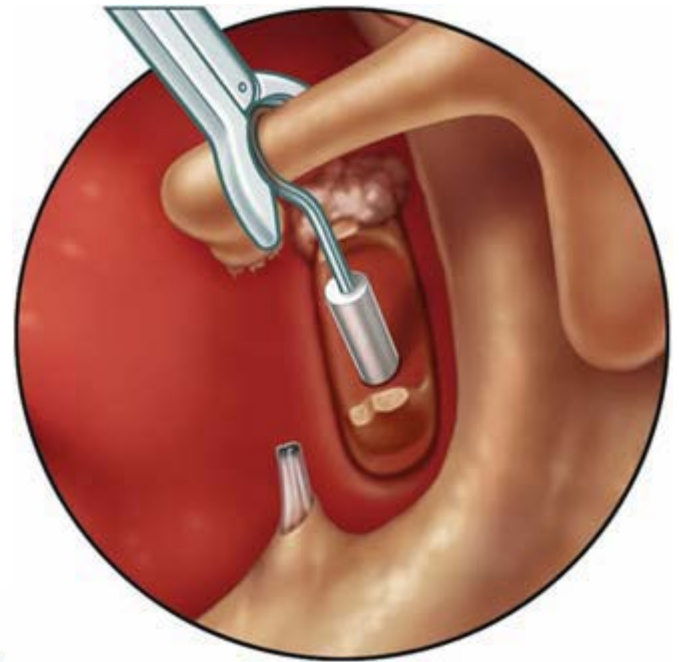
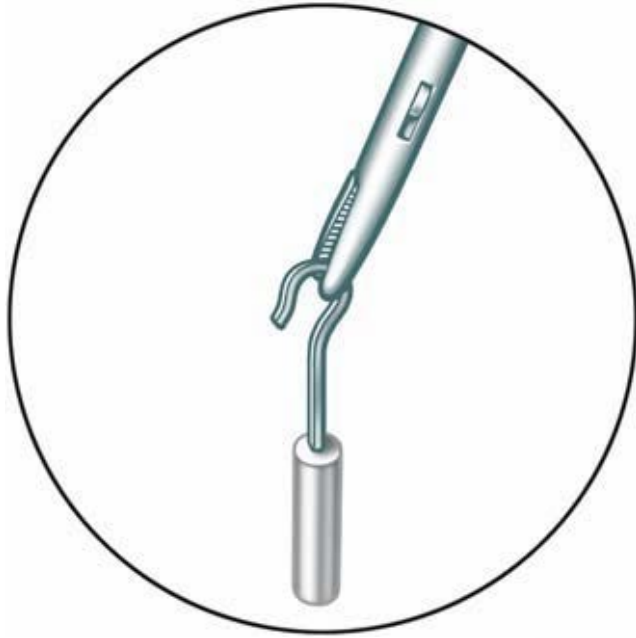


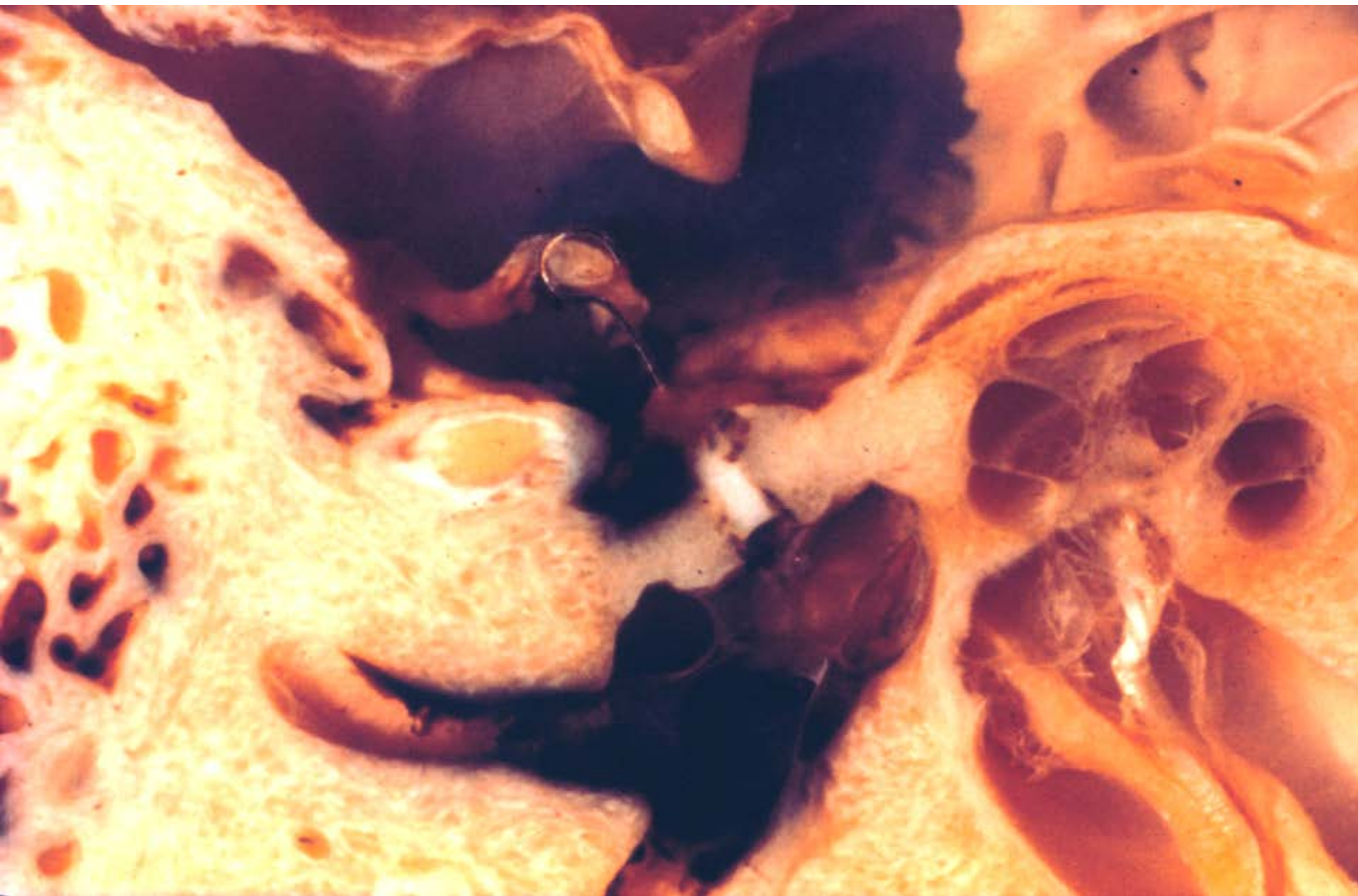
Decision to plan stapes surgery without CT scan in this case

Next question: plan the surgery in local or general anesthesia?









Stapes surgery in local or general anesthesia?

General vs. local anesthesia in stapes surgery

- No evidence for advantage for one over the other in terms of audiological results
- Up to the patient and the surgeon to find a suitable solution for the individual case

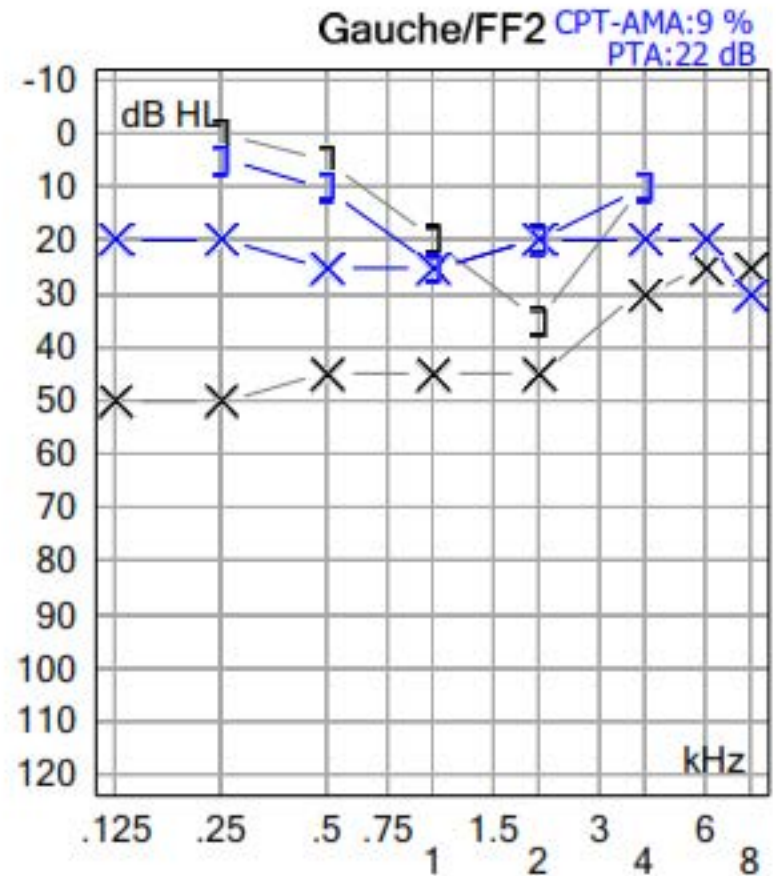
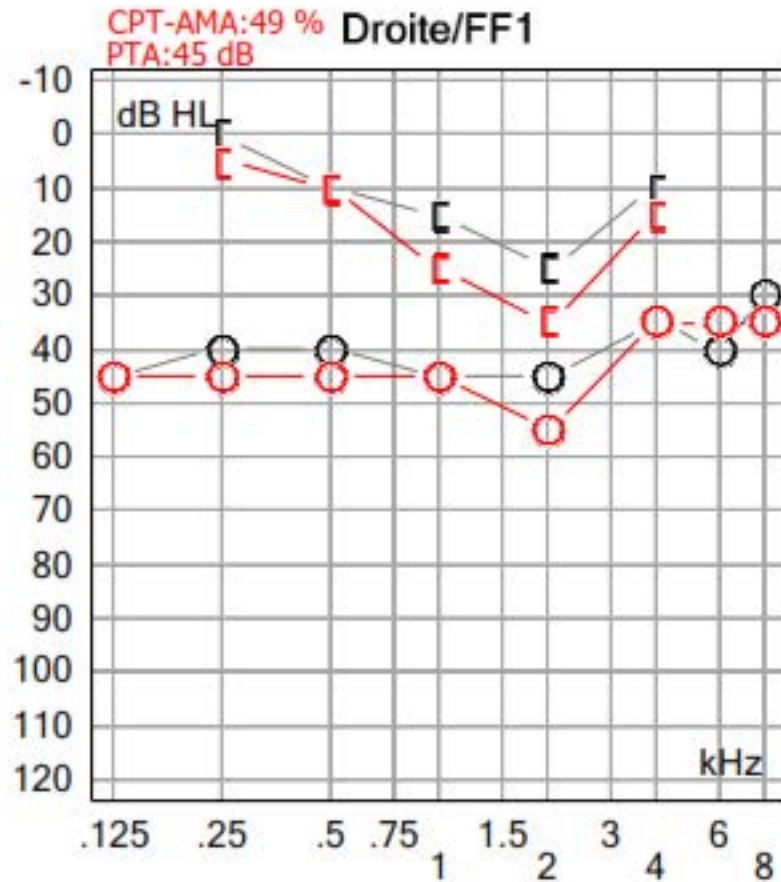
Otolaryngol Head Neck Surg. 2013 Dec;149(6):963.

RE: Local versus general anesthesia for stapes surgery. Corrales CE, Jackler RK.

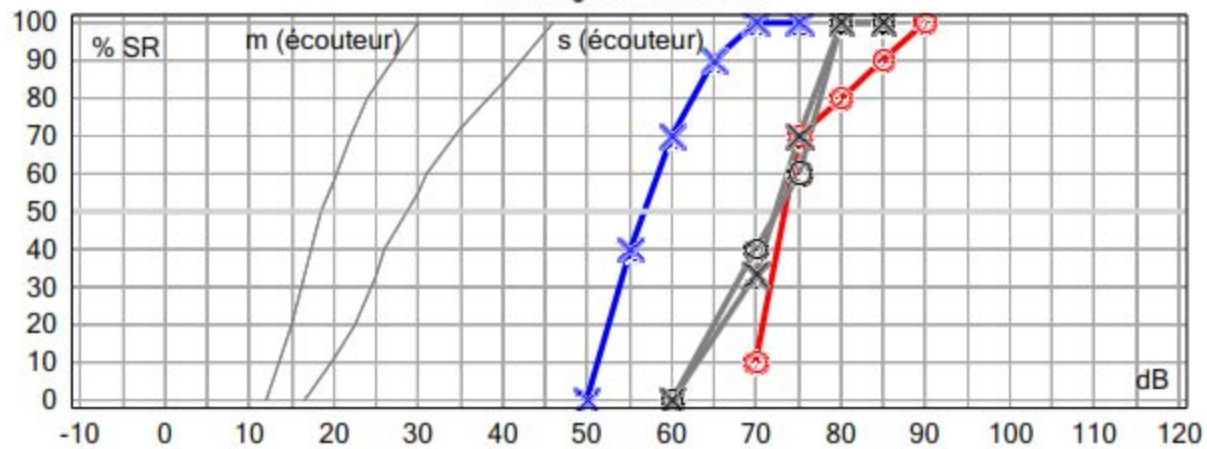
Otolaryngol Head Neck Surg. 2013 Sep;149(3):360-5. Local versus general anesthesia in stapes surgery for otosclerosis: a systematic review of the evidence. Wegner I1, Bittermann AJ, Zinsmeister MM, van der Heijden GJ, Grolman W.

VIDEOS AND FOTOS FROM THE SURGERY

AUD 16.08.2016
AUD 04.12.2015



Audiogramme vocal



Laser stapedotomy film

Conventional vs. Laser stapedotomy

- The majority of studies hints at less complications (bleedings and sensorineural hearing loss) for laser stapedotomy¹
- Fewer studies show equal results and complication rates for both procedures²

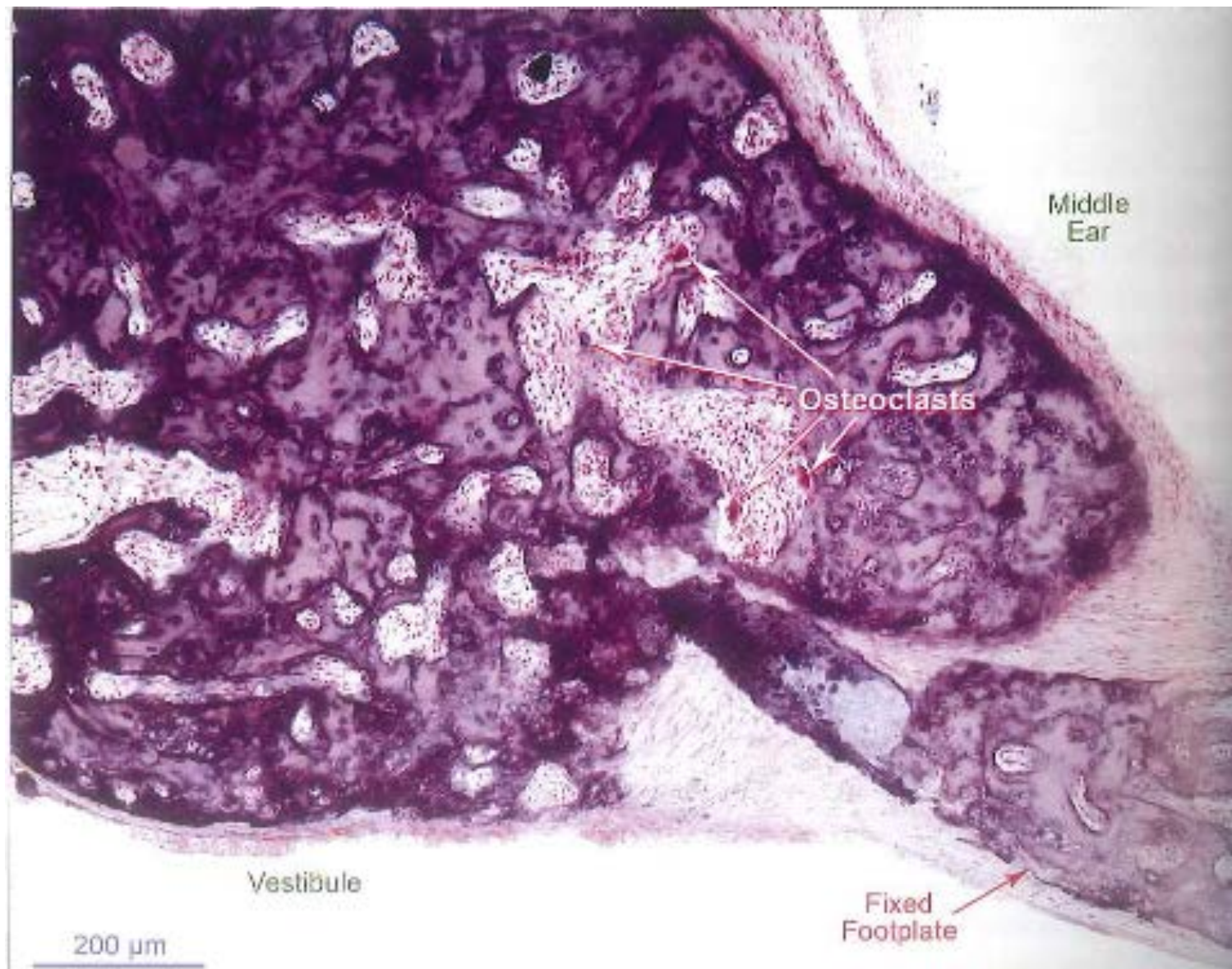
¹ Laryngoscope. 2014 Jul;124(7):1687-93. Laser versus conventional fenestration in stapedotomy for otosclerosis: a systematic review. Wegner L, Kamalski DM, Tange RA, Vincent R, Stegeman I, van der Heijden GJ, Grolman W.

² Otolaryngol Head Neck Surg. 2016 Jun;154(6):1099-105. doi: Diode Laser Stapedotomy vs Conventional Stapedotomy in Otosclerosis: A Double-Blinded Randomized Clinical Trial. Parida PK, Kalaiarasi R, Gopalakrishnan S.

Otosclerosis definition¹

- Bone remodeling disease of the enchondral bone with 4 histological hallmarks
 - Decalcification
 - Hypervascularization
 - Fibrosis
 - Neo-osteogenesis
- Incidence of non-clinical otosclerosis sites in histological studies 8 – 13% in Caucasians
- In clinical otosclerosis cases, 96% have **fenestral** and 49% additional **cochlear** sites

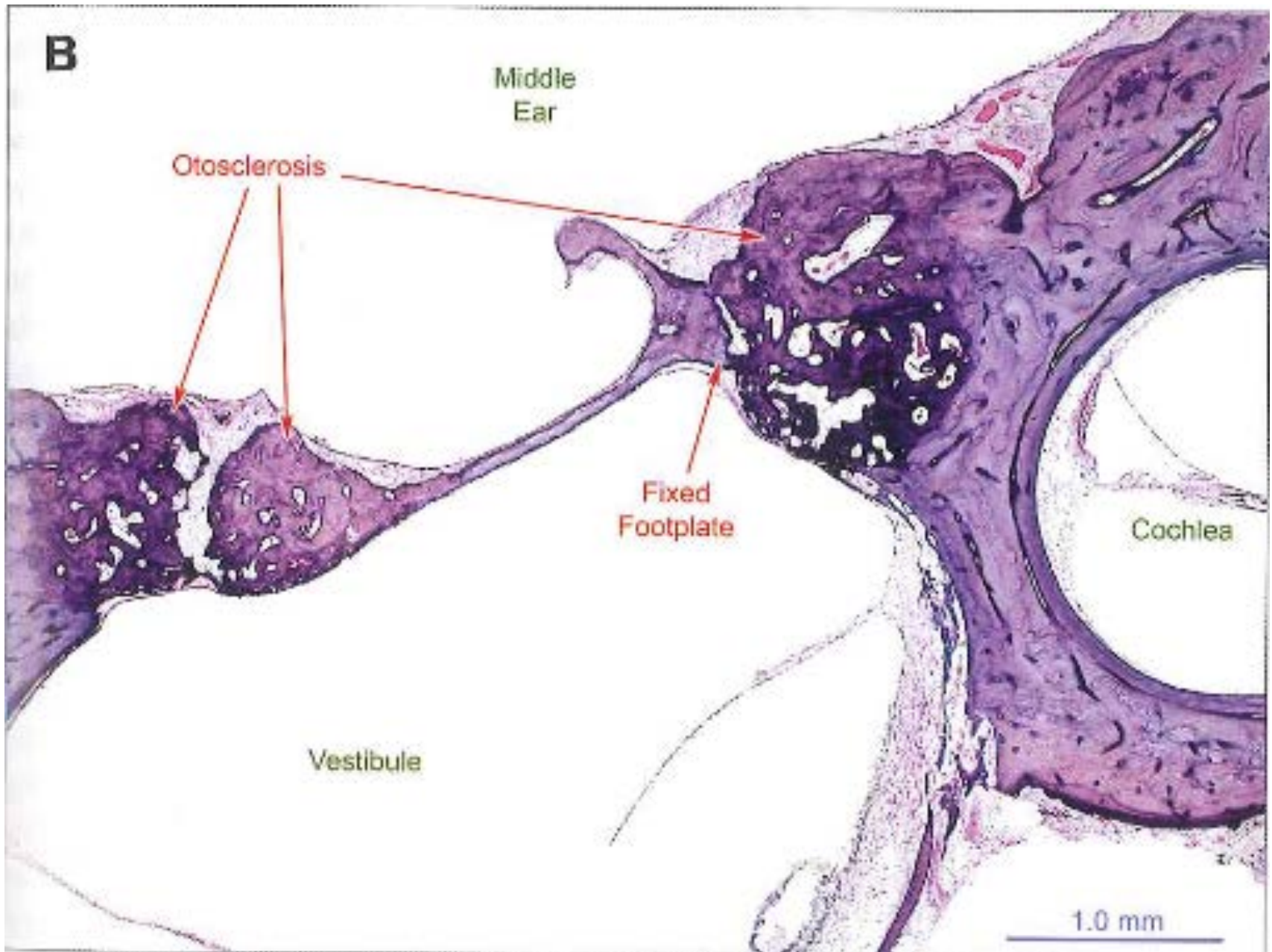
¹Schuknecht's Pathology of the Ear, 3rd edition, 2010



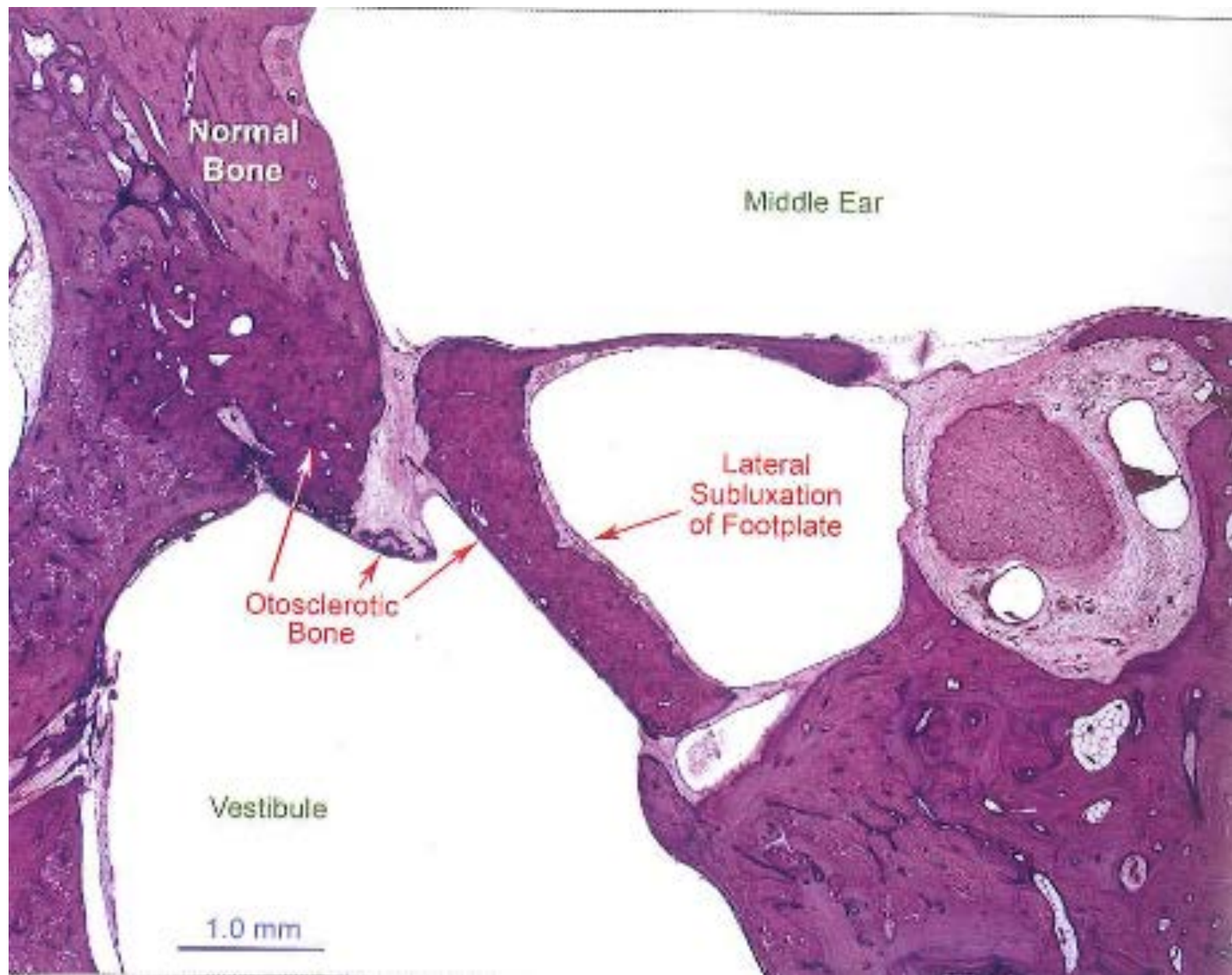
Clinial presentation

- Progressive hearing loss without history for trauma or ear infections
- Family history positive in about 50%
- Vertigo and imbalance in some patients
- Progression in pregnancy in some women
- **Conductive** or mixed hearing loss with intact tympanic membrane, missing stapedial reflex and in some cases red shining promontory (Schwartz sign)

Fenestral Otosclerosis







Otosclerosis facts

- Clinical prevalence 1% in Caucasians^{1,2}
- Etiology: combination of genes and environment³
 - Probably autosomal dominant inheritance with incomplete penetrance²
 - >20 genes associated with otosclerosis²
 - measles virus infection possible³

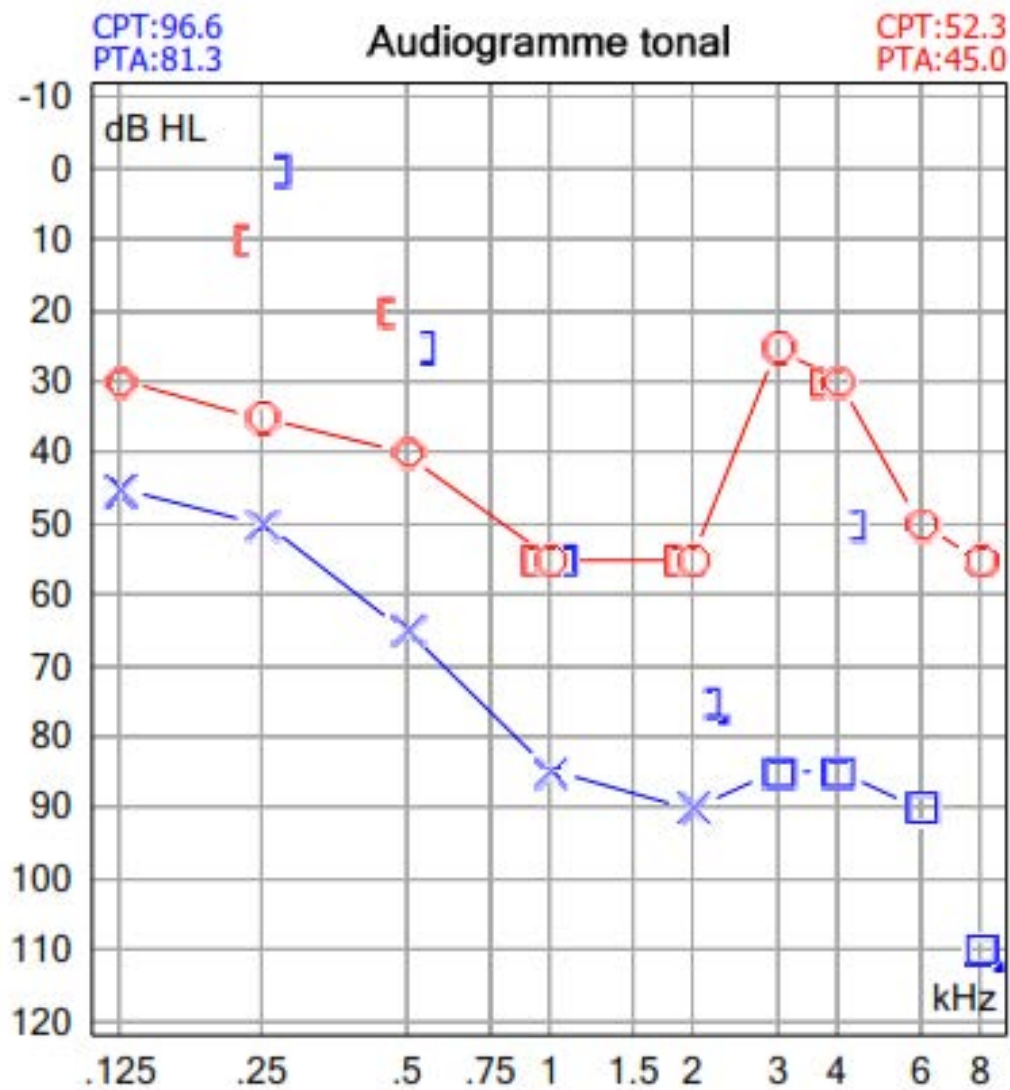
¹Hannula S et al. Ear diseases and other risk factors for hearing impairment among adults: an epidemiological study. Int J Audiol. 2012 Nov;51(11):833-40

²Schuknecht's Pathology of the Ear, 3rd edition, 2010

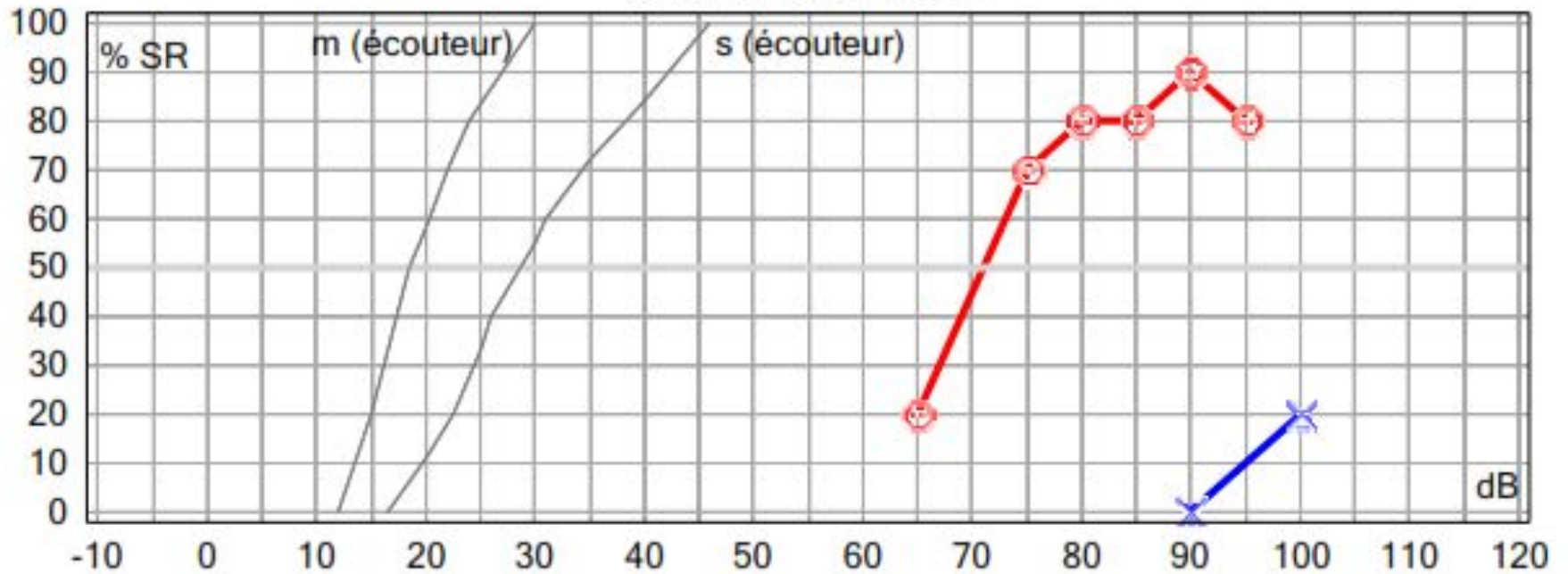
³Schrauwen I, Van Camp G. The etiology of otosclerosis: a combination of genes and environment. Laryngoscope. 2010 Jun;120(6):1195-202.

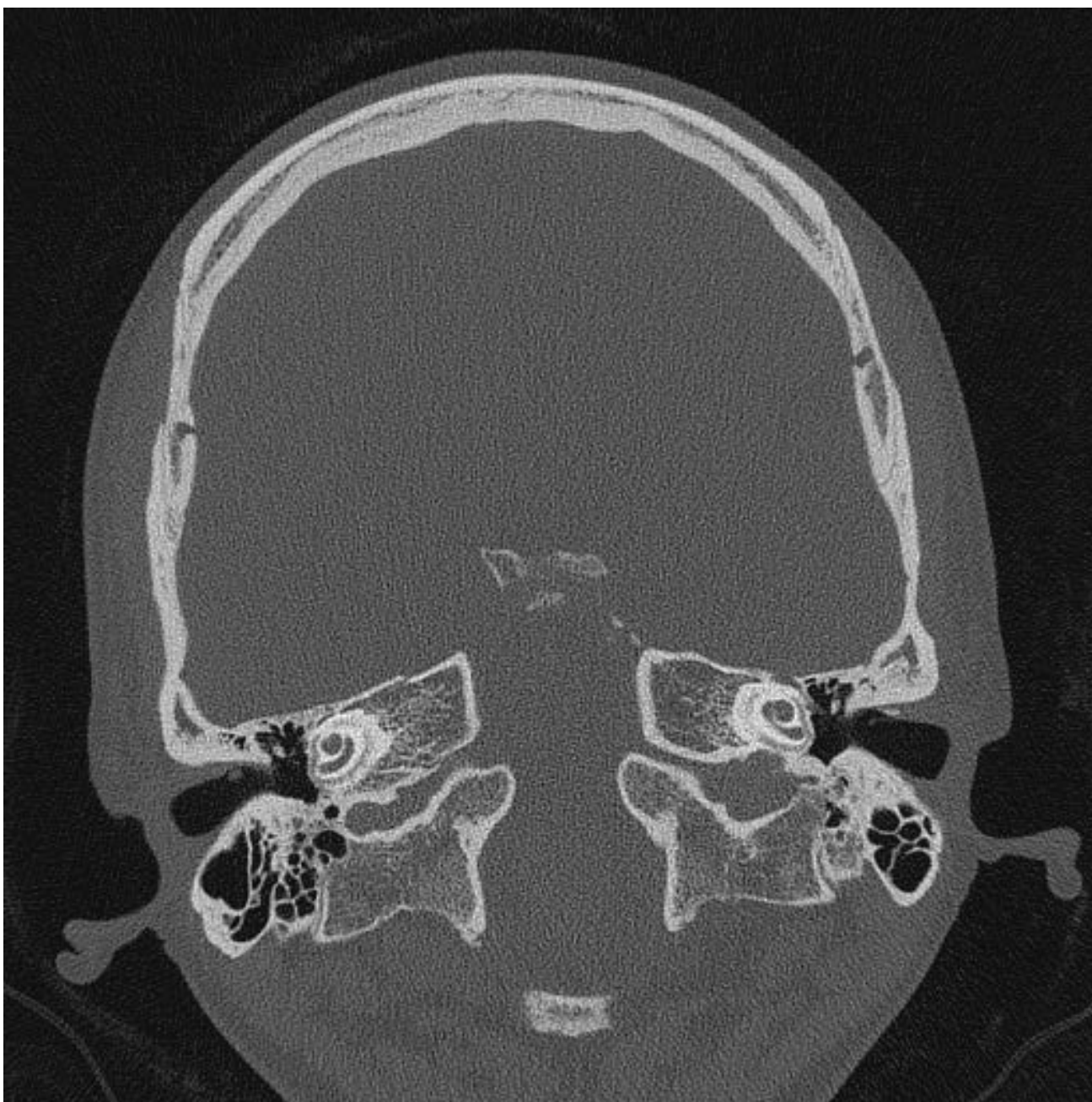
42-year old fashion designer

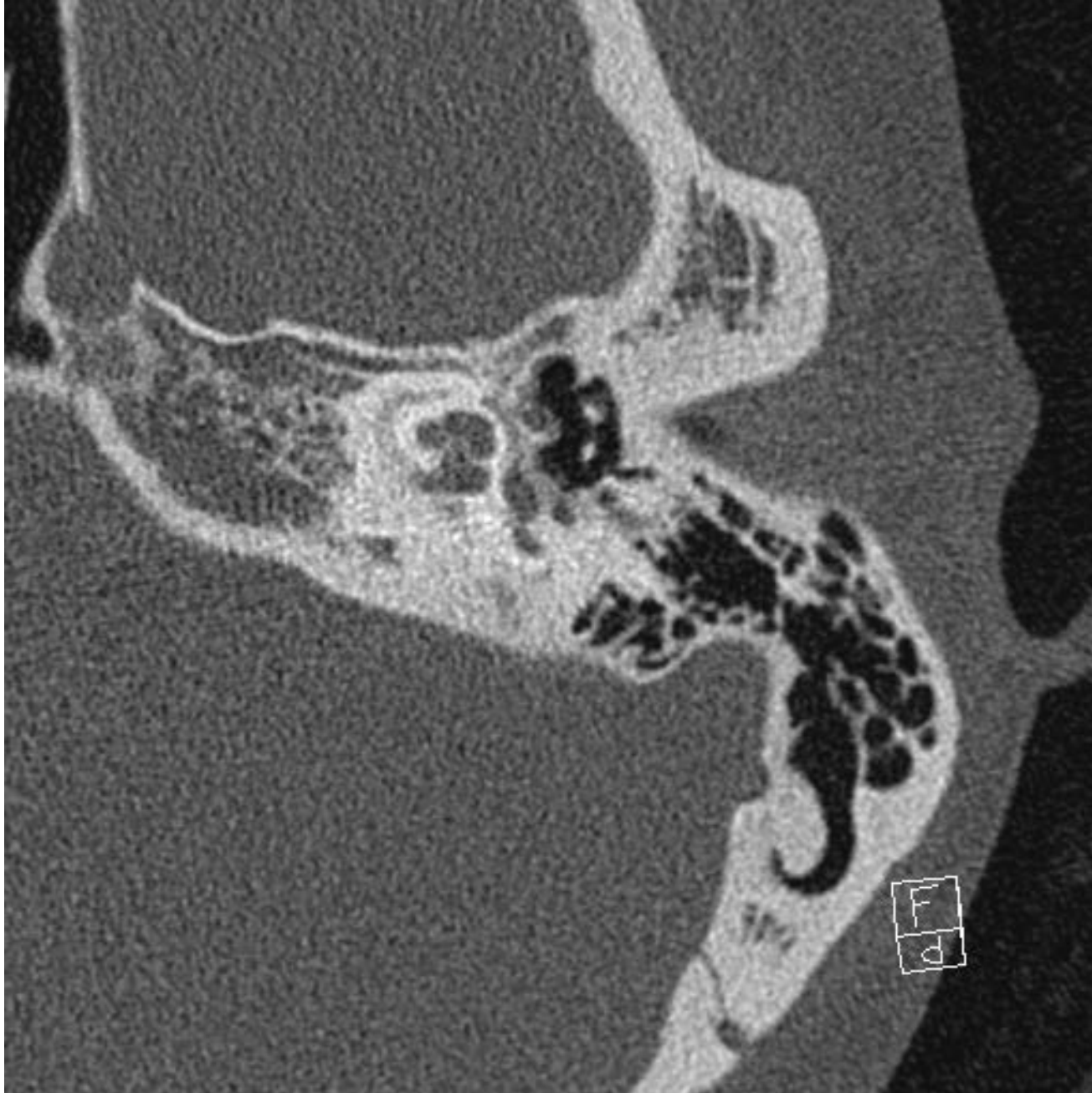
- Progressive, bilateral hearing loss since many years without other symptoms
- Negative family history for hearing loss
- Status: Weber centered/right, Rinne négatif bilatéralement, membranes tympaniques intactes avec signe de Schwartze bilatéralement

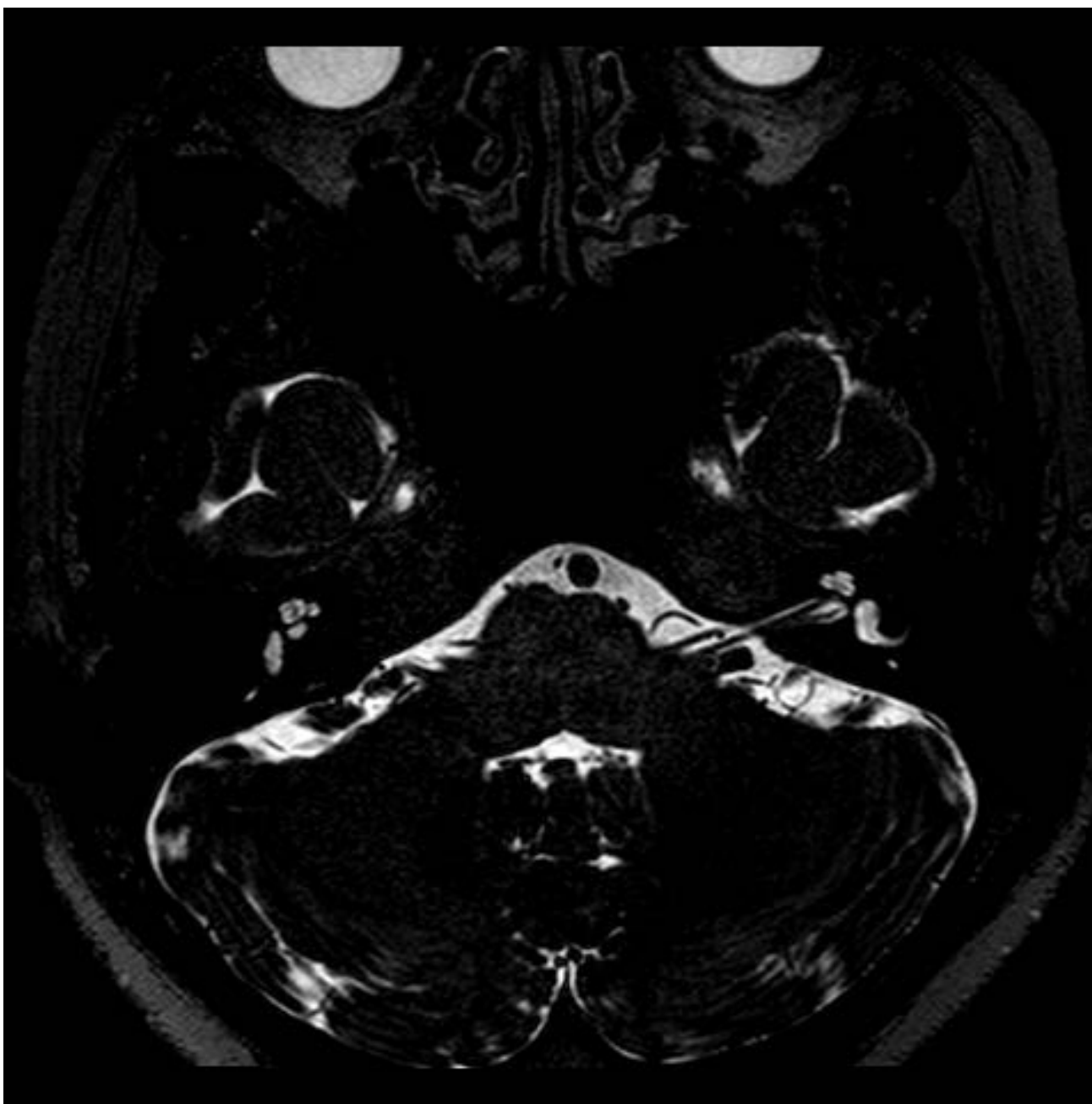


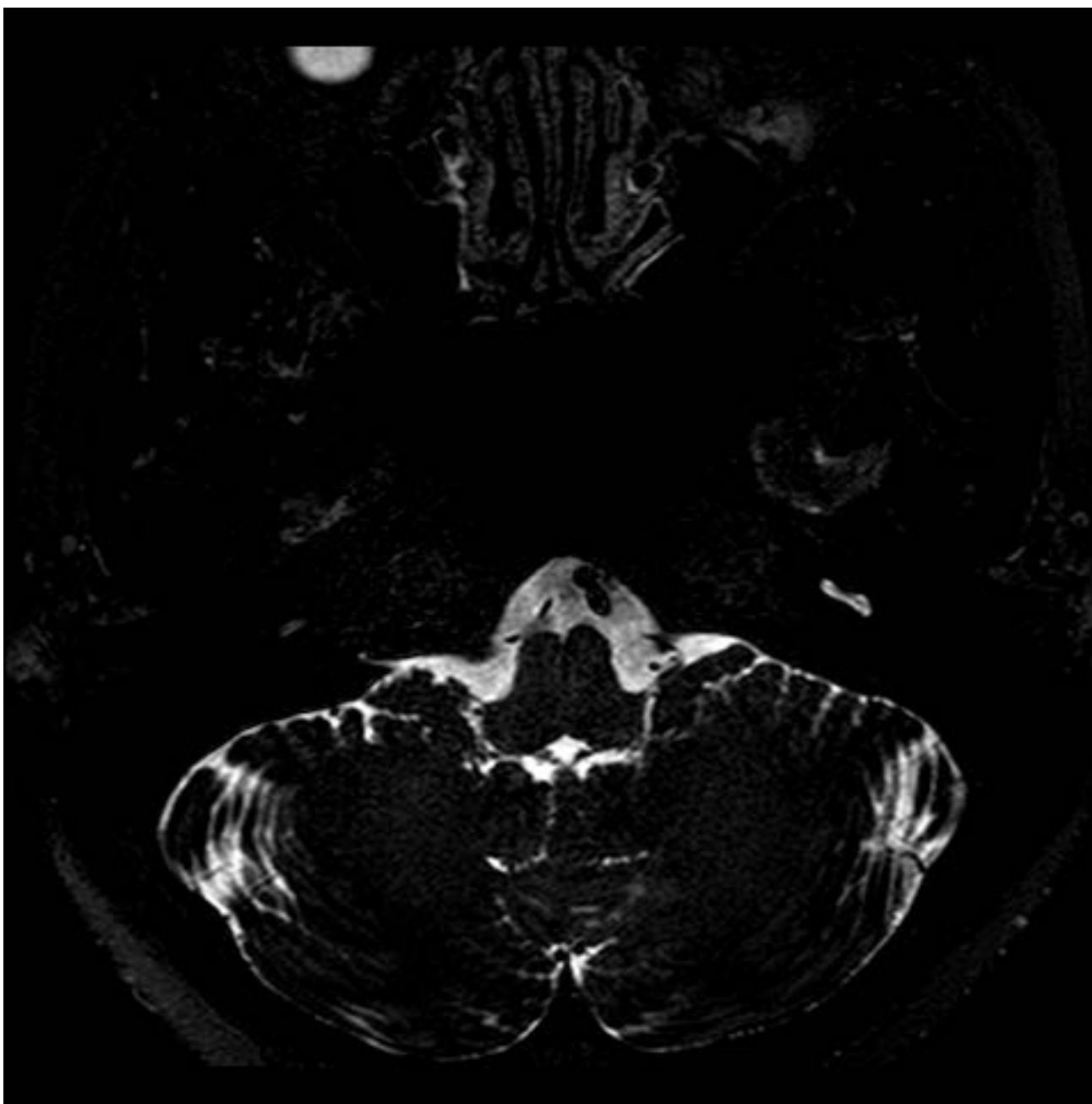
Audiogramme vocal





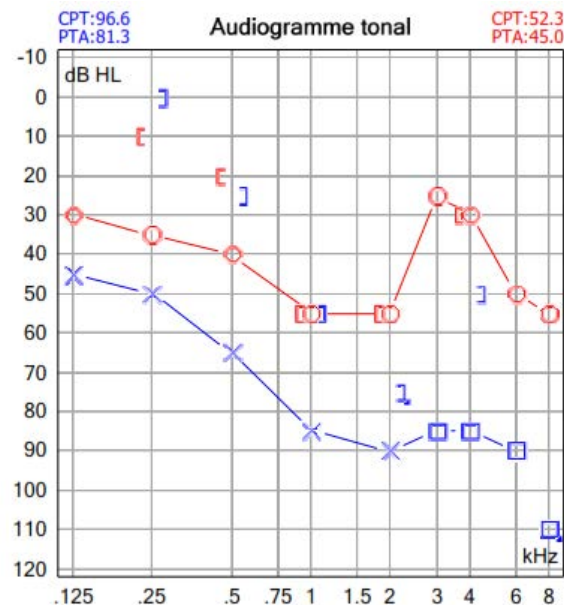




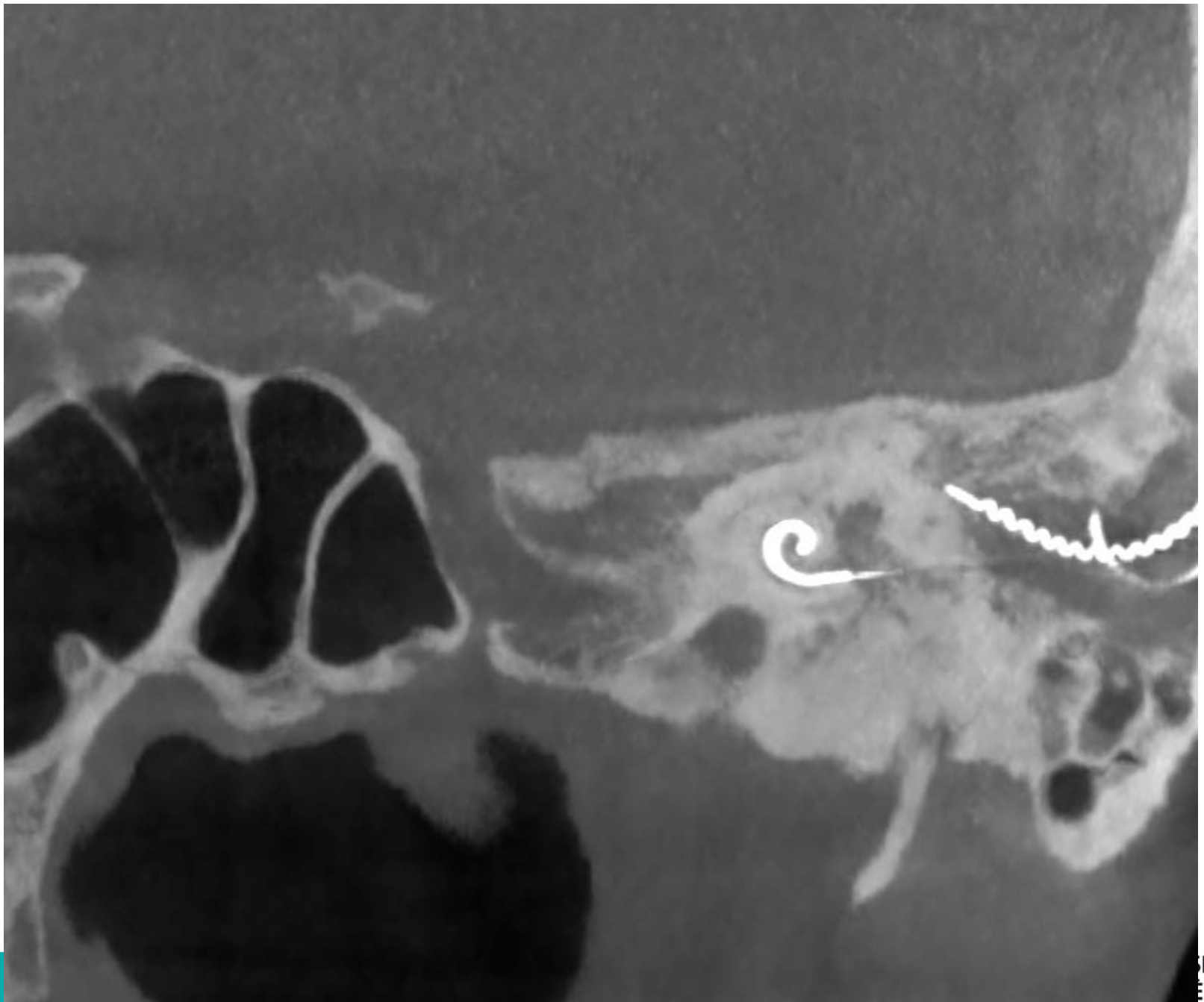


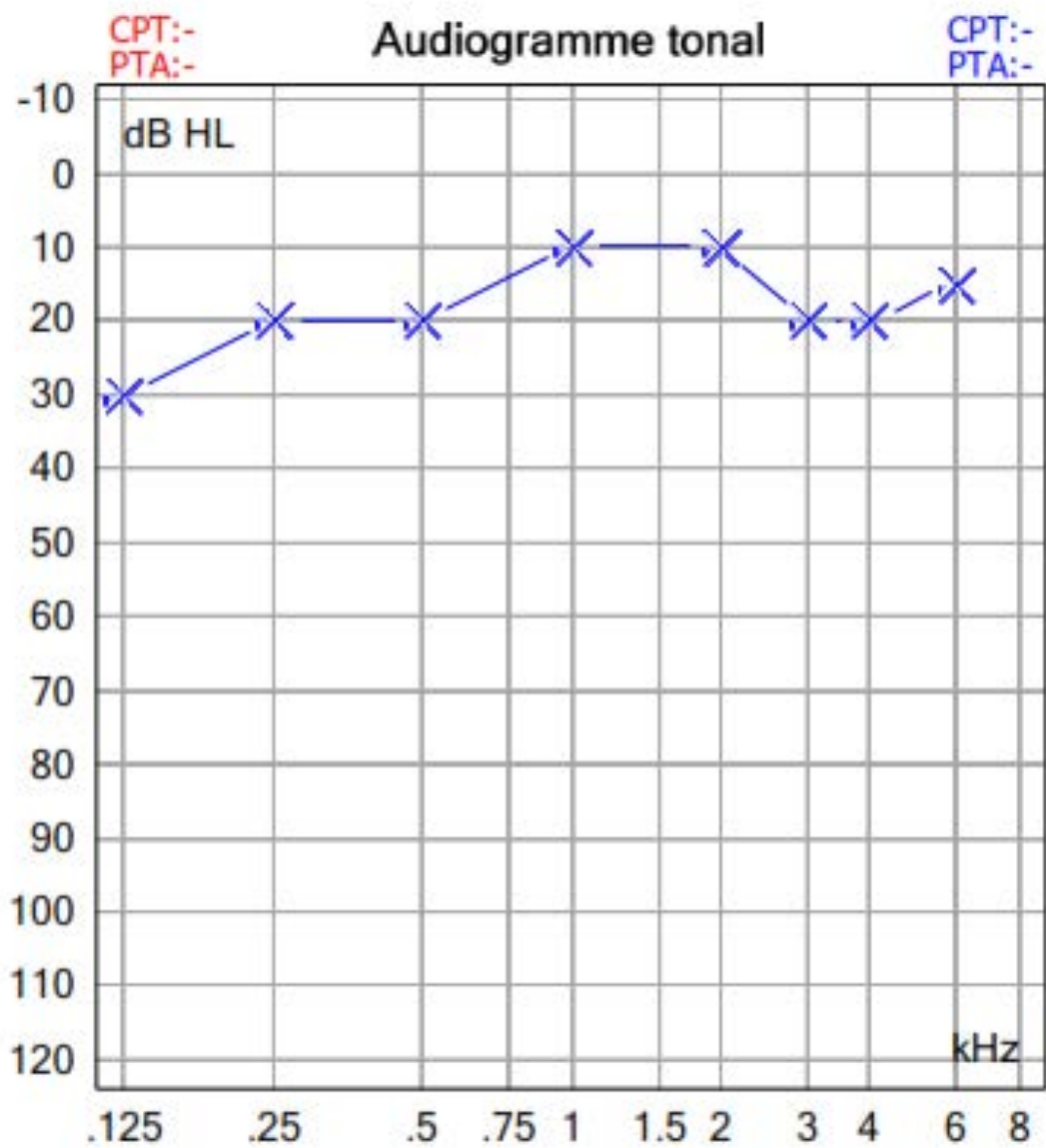
42-year old fashion designer

- Severe, mixed hearing loss on the left side without stapedial reflex and confirmed cochlear otosclerosis
- Moderate, mixed hearing loss on the right side



What therapy to propose?



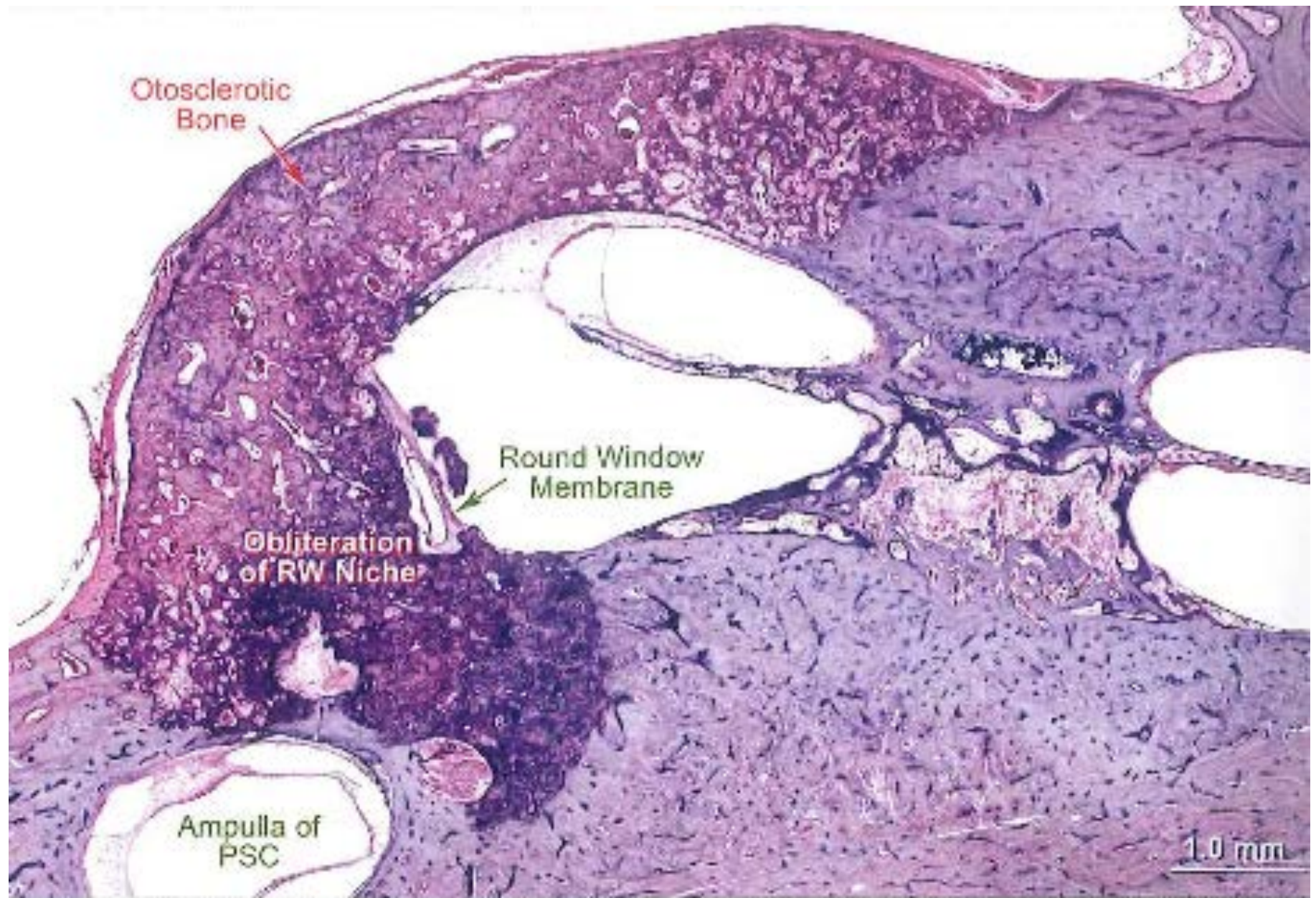


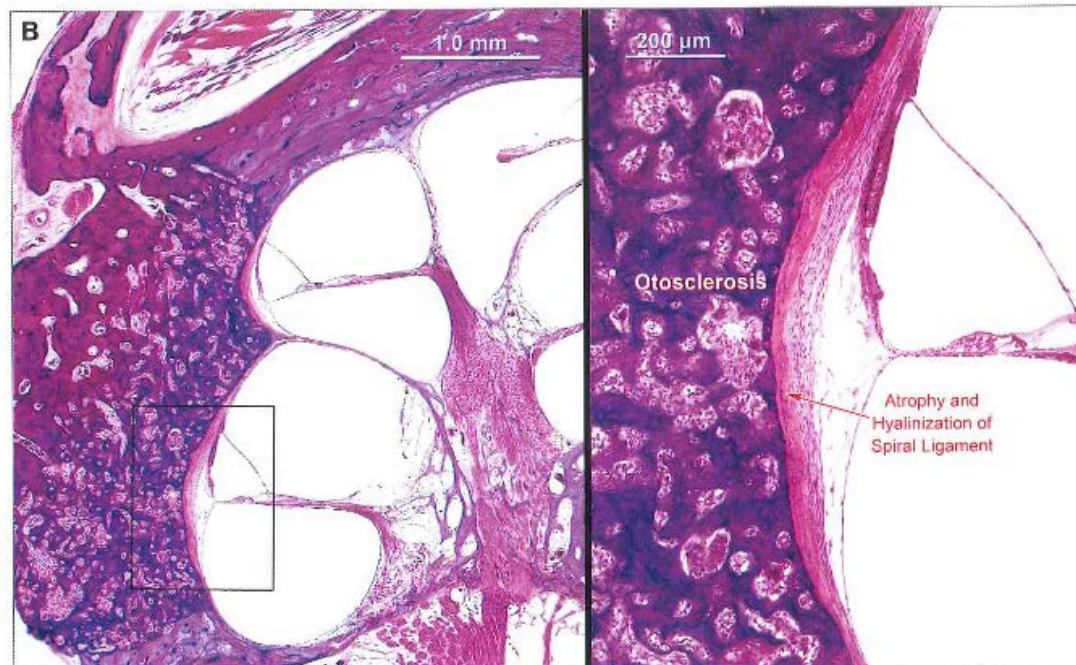
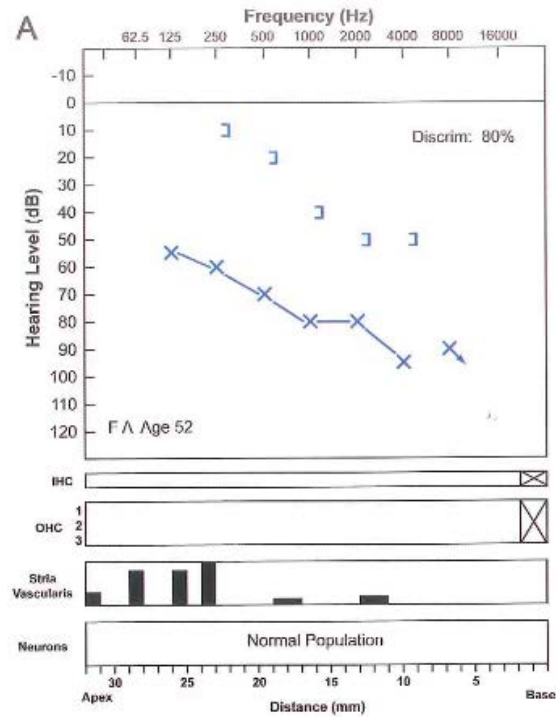
Cochlear otosclerosis

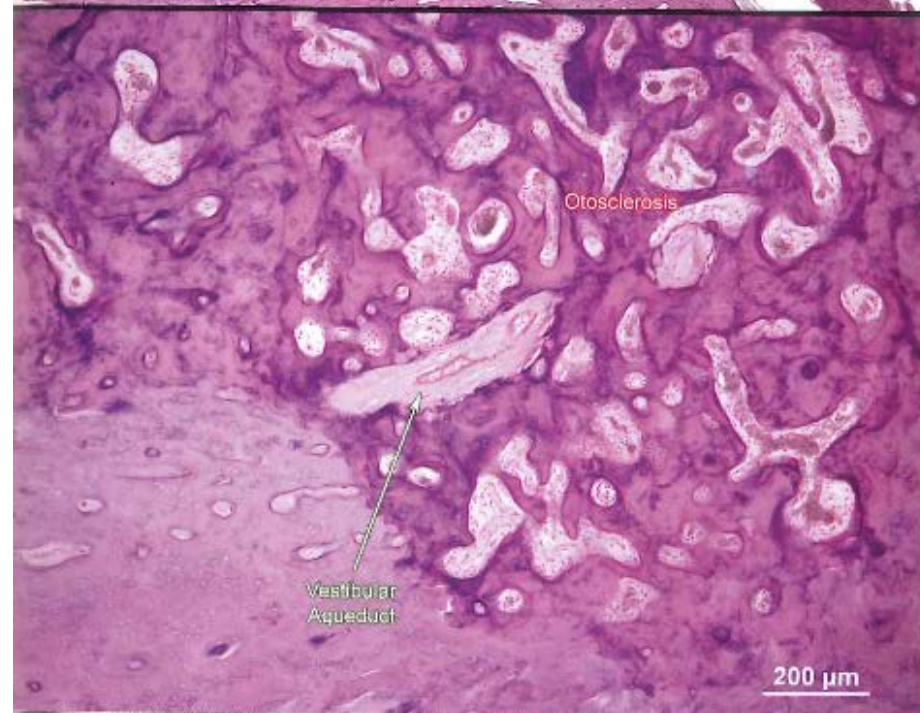
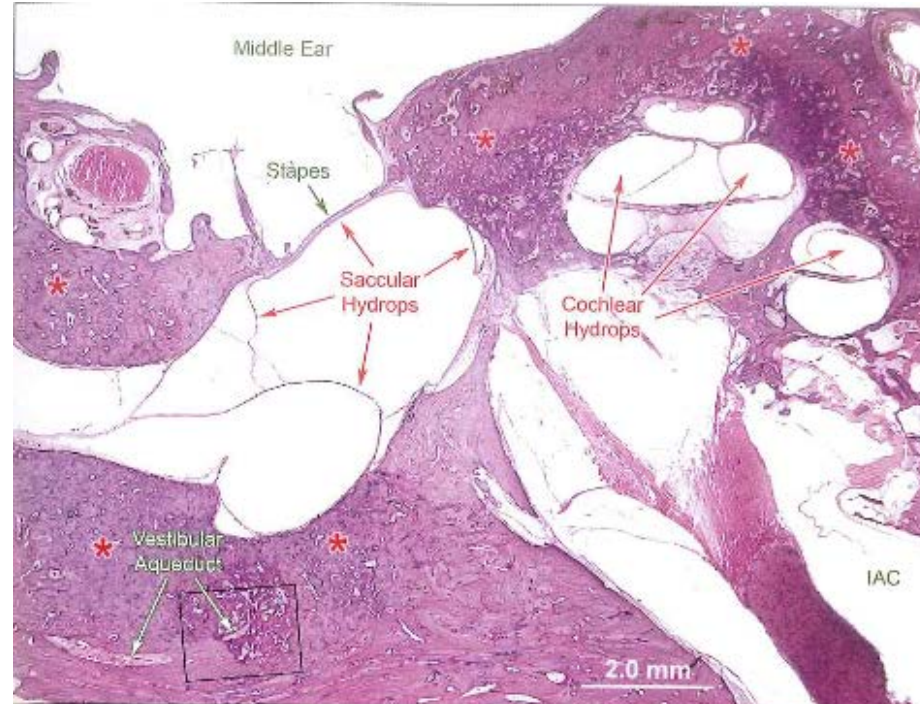
- Histologically (not clinically) present in 49% of all otosclerosis cases¹
- **Sensorineural hearing loss starts, when endosteum involved – MIXED hearing loss (stapes fixed)**
- Otosclerotic lesions around the cochlea with atrophy of stria vascularis
 - Vestibular aqueduct affected: endolymphatic hydrops with Menière symptoms
- Therapeutic considerations: Stapedotomy vs. Hearing aid vs. Combination vs. Cochlear implantation²

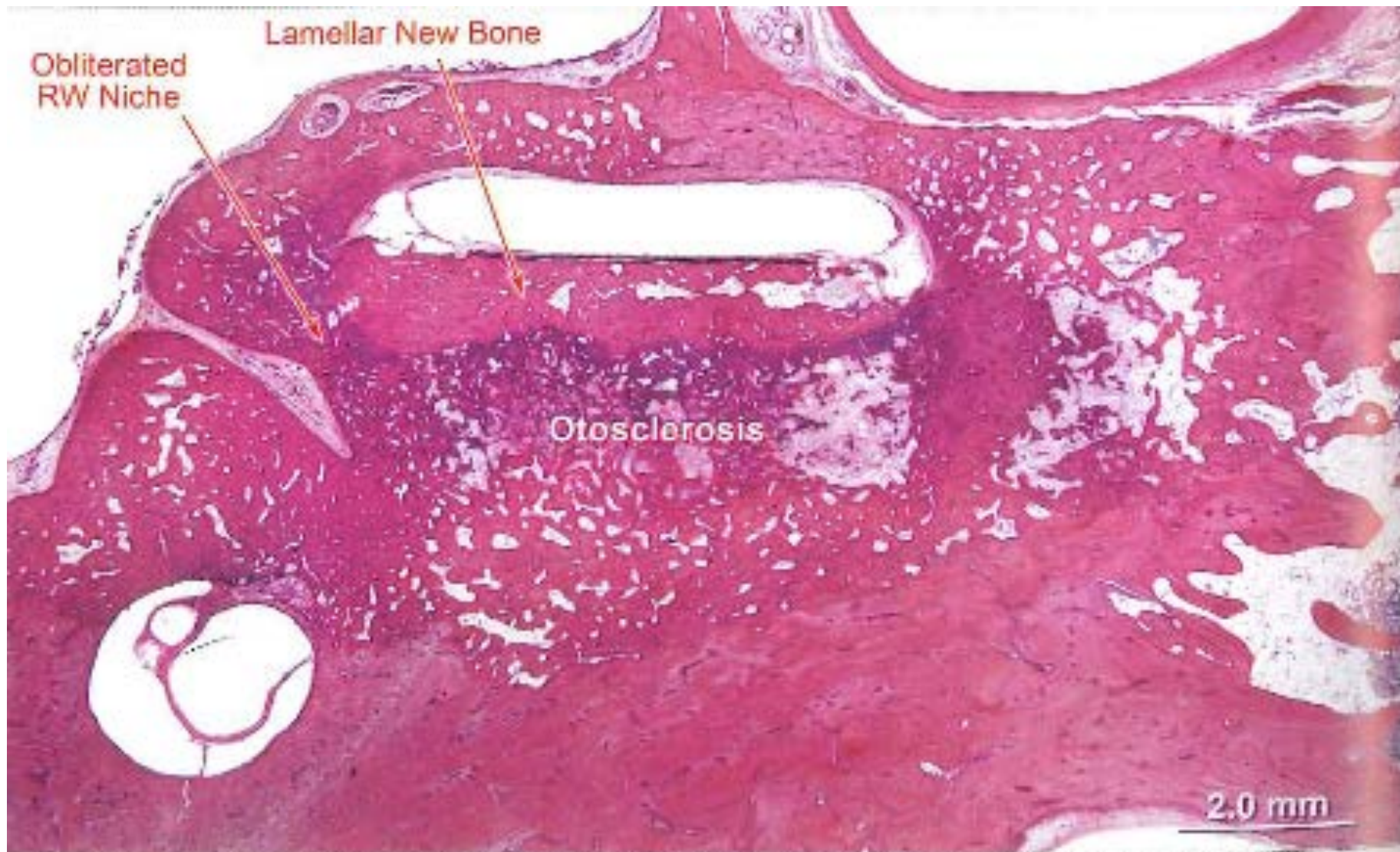
¹Schuknecht's Pathology of the Ear, 3rd edition, 2010

²Markus P et al. Laryngoscope. 2011 Sep;121(9):1935-41. Decision making in advanced otosclerosis: an evidence-based strategy.









Non-surgical therapeutic options

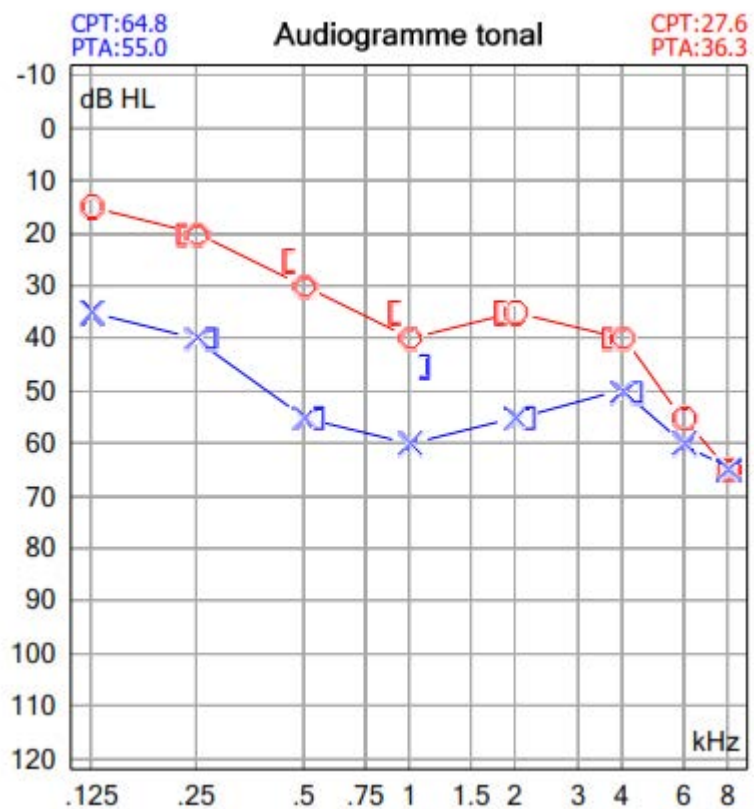
- Hearing aids (no risk)
 - Bone anchored hearing aids (BAHA)
 - Conventional hearing aids
- Medical treatment to slow down bone remodeling process in cases with sensorineural hearing loss
 - Bisphosphonates¹
 - Systemic¹
 - Local² (preclinical animal models)

¹Quesnel AM et al. Otol Neurotol. 2012 Oct;33(8):1308-14. Third-generation bisphosphonates for treatment of sensorineural hearing loss in otosclerosis.

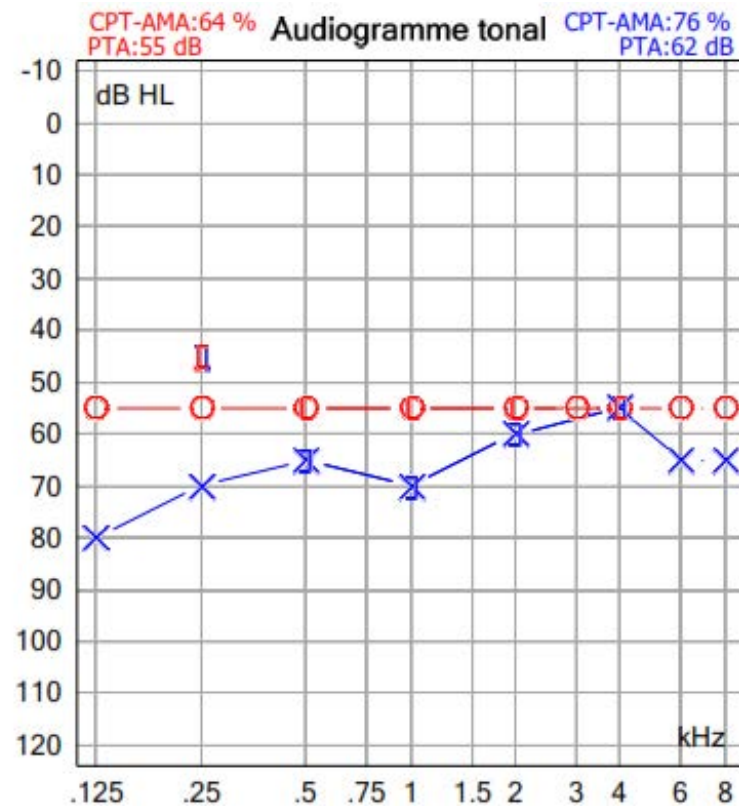
²Kang WS et al. Otol Neurotol. 2015 Jul;36(6):953-60. Non-Ototoxic Local Delivery of Bisphosphonate to the Mammalian Cochlea.

34-year old office employee

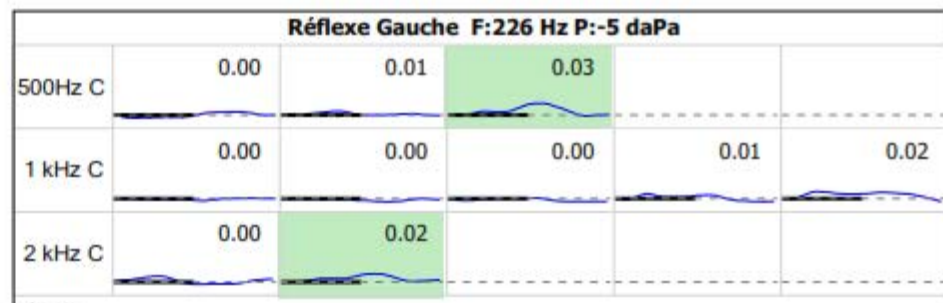
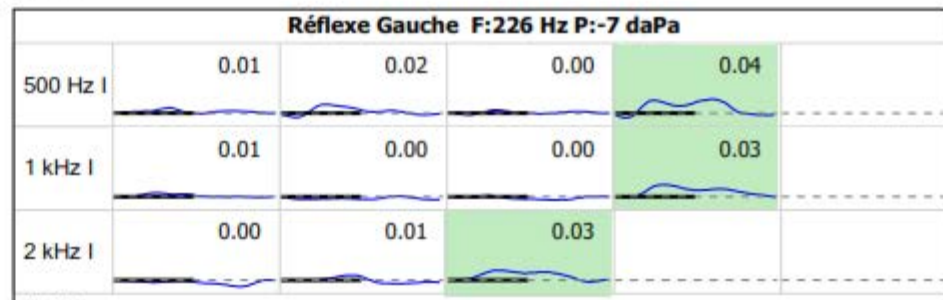
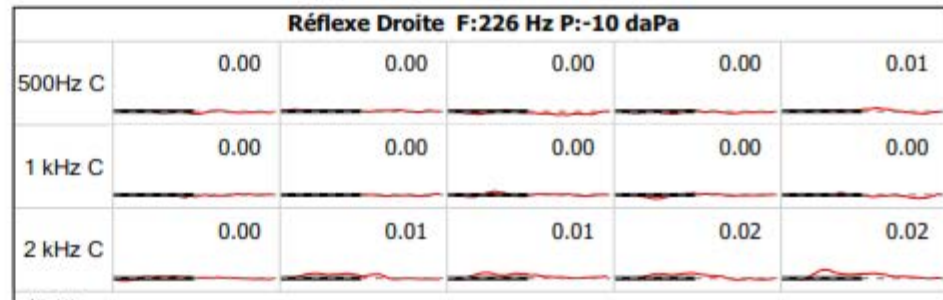
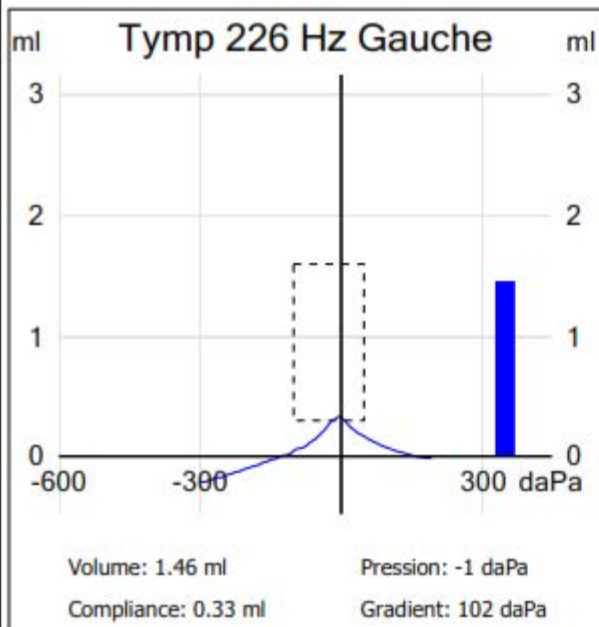
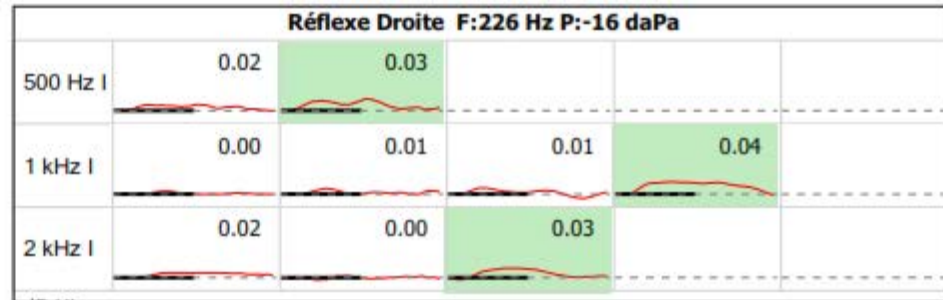
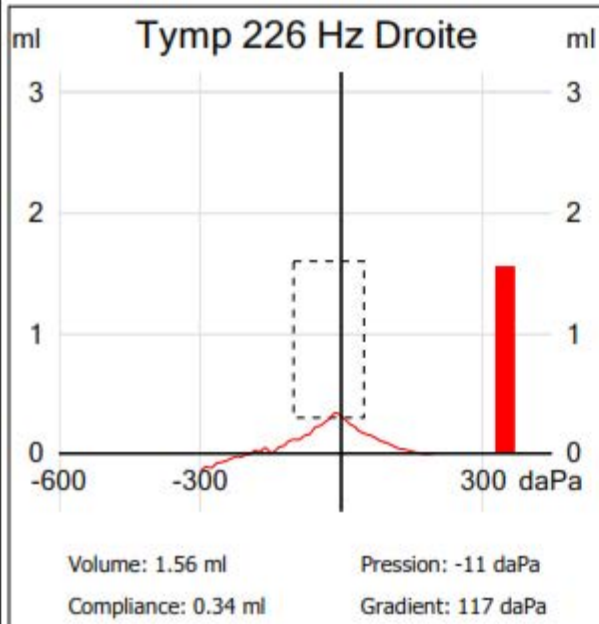
- Non-familial, bilateral progressive hearing loss since about 7 years with at least 2 episodes of vertigo and imbalance
- Wears hearing aids bilaterally since 5 years
- Status: Weber centered, Rinne **positif** bilaterally, intact tympanic membranes with Schwartz sign

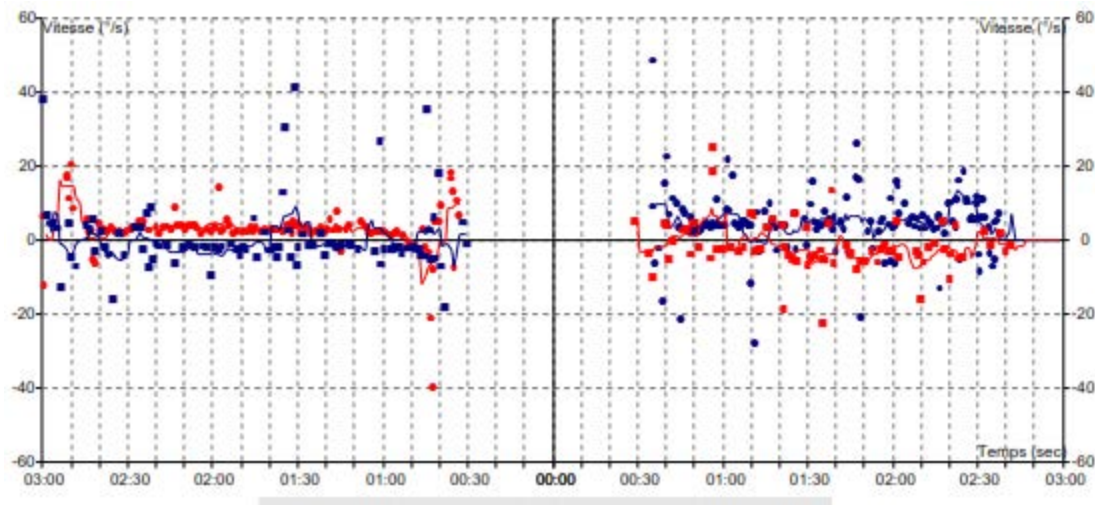


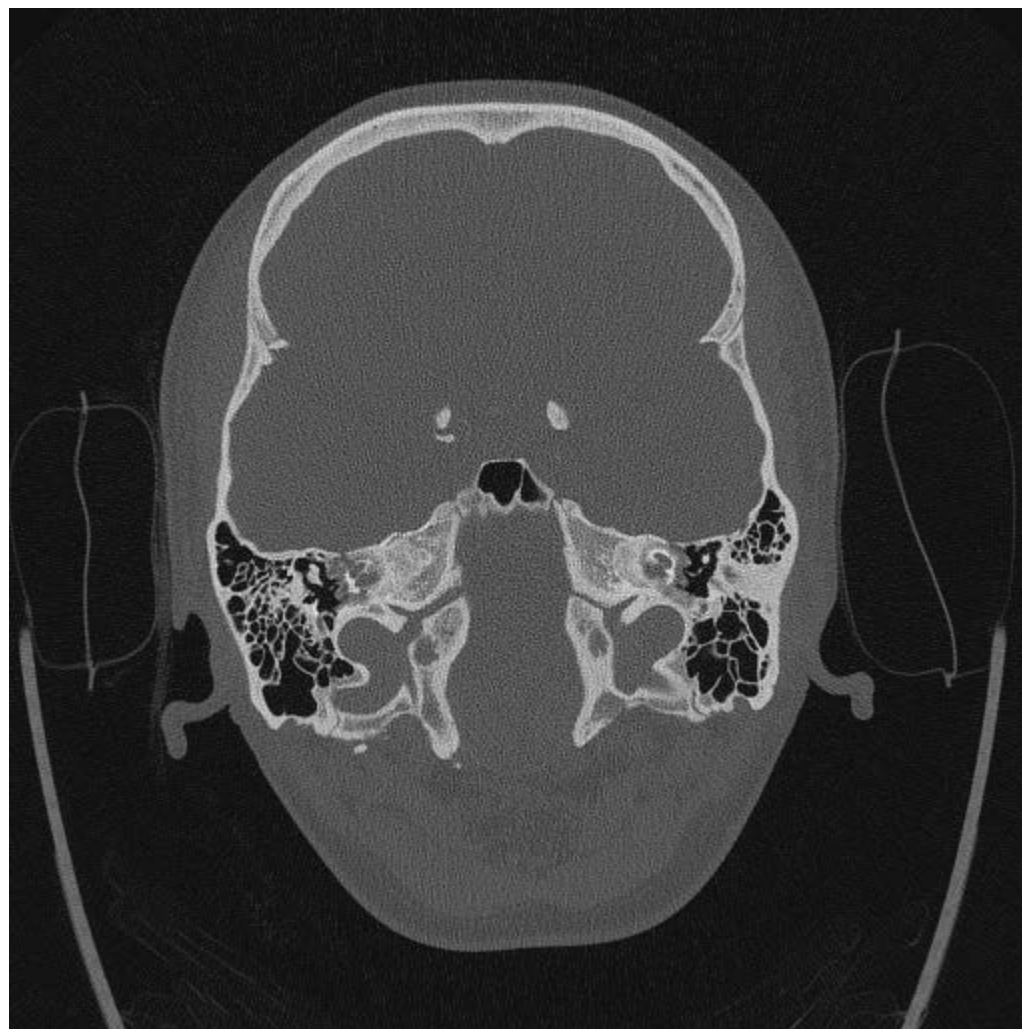
2013

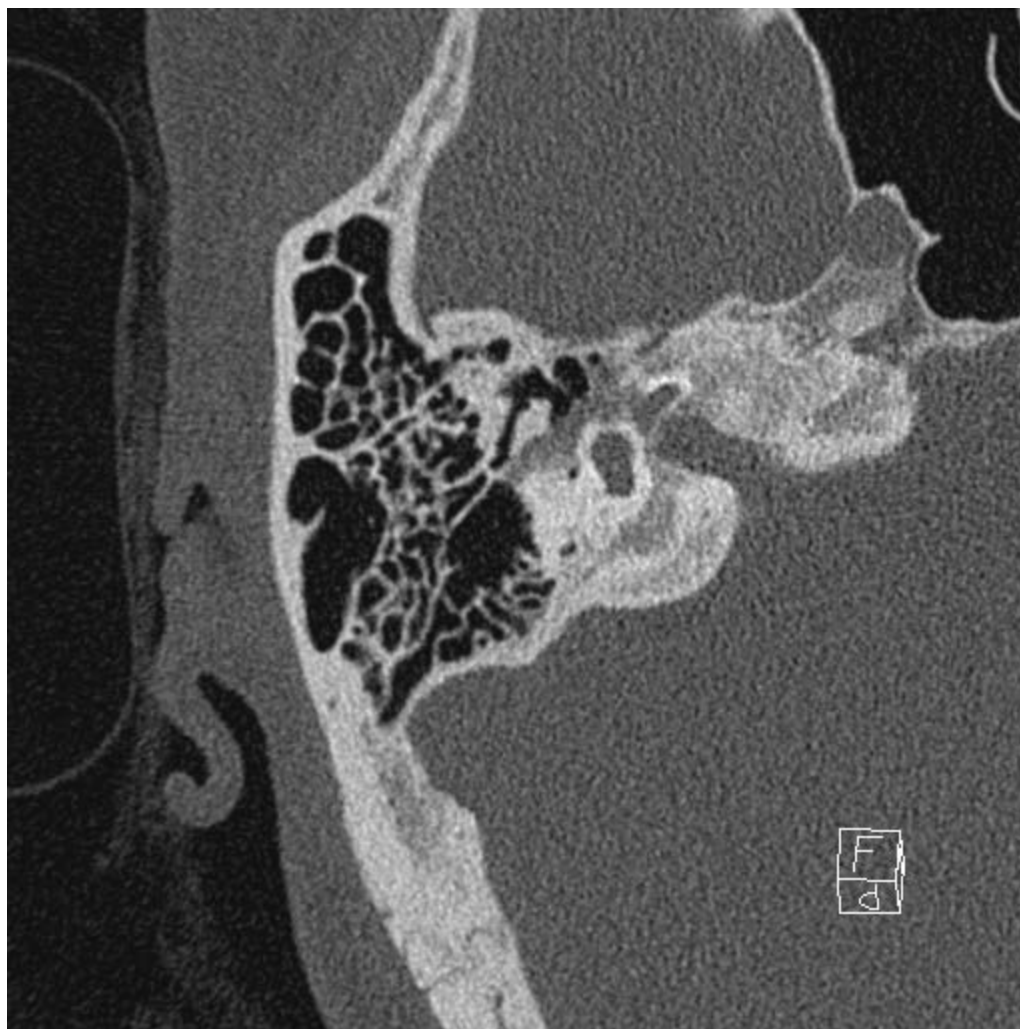


2015

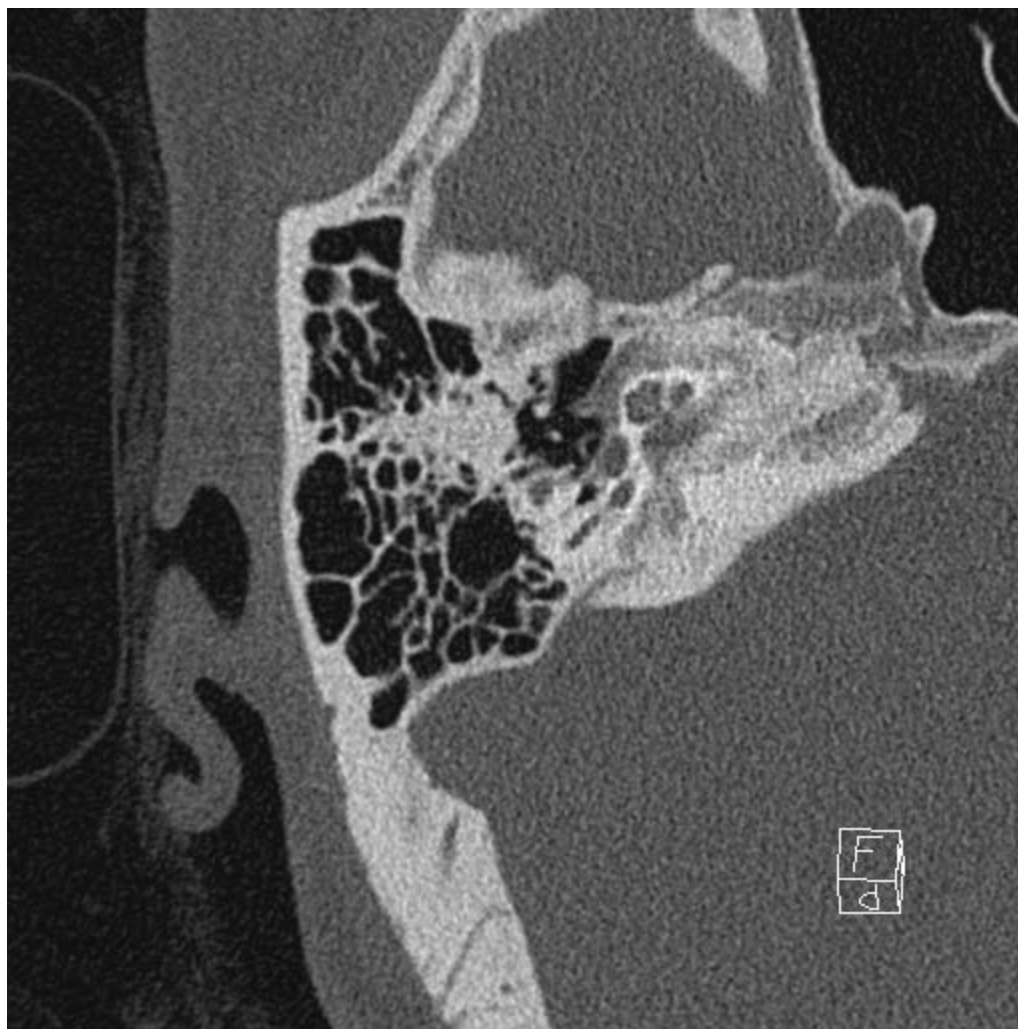






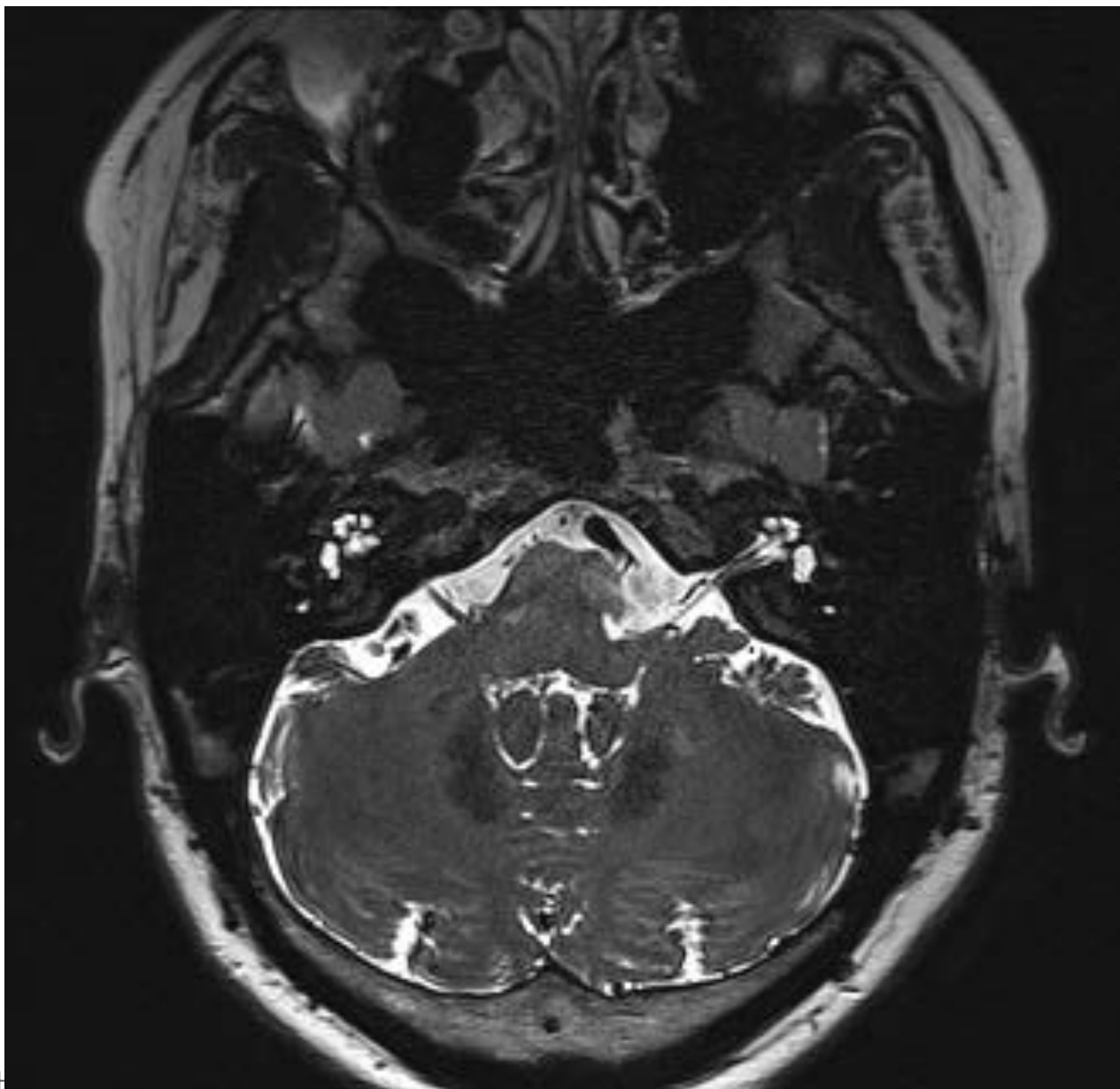


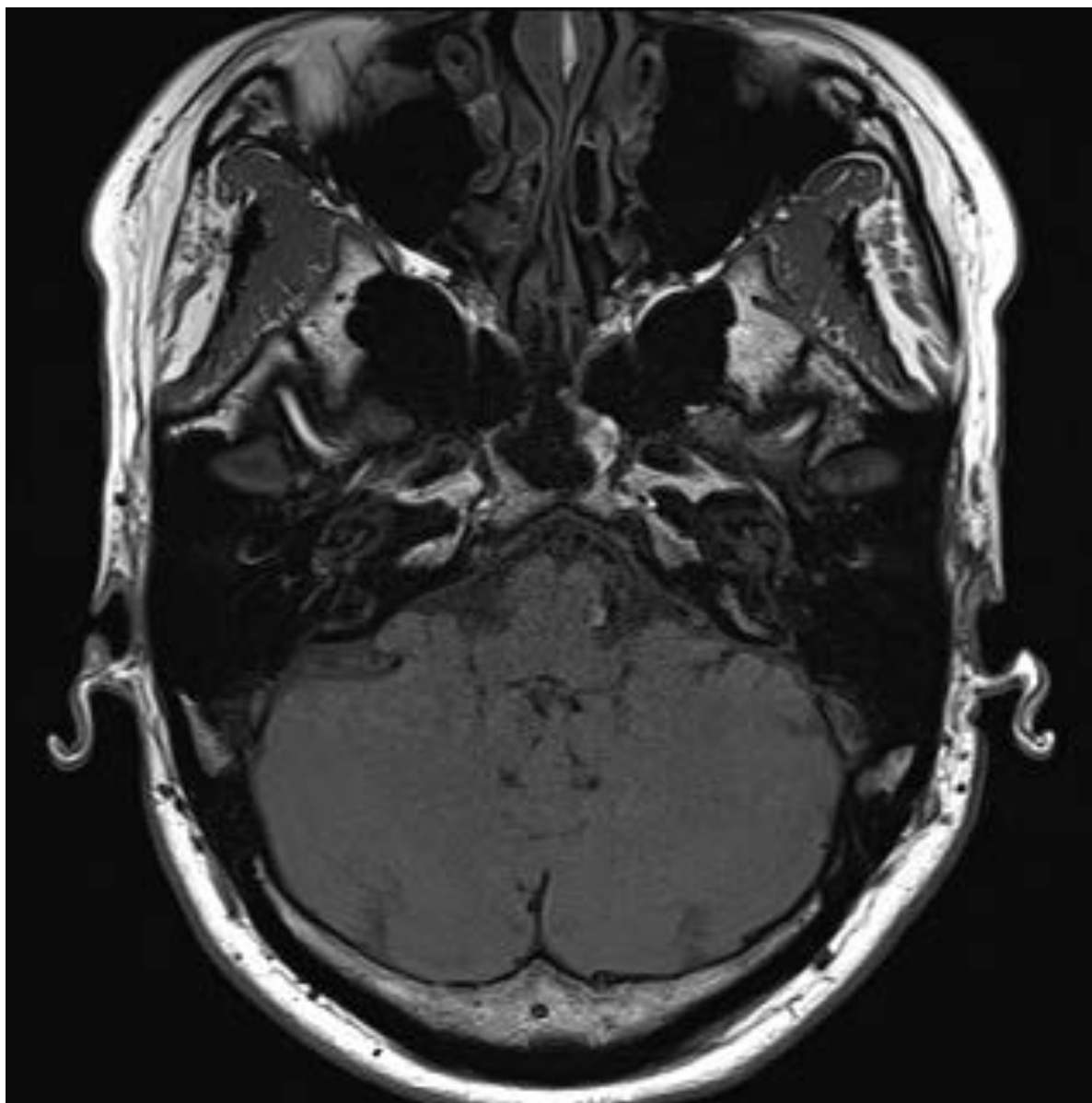


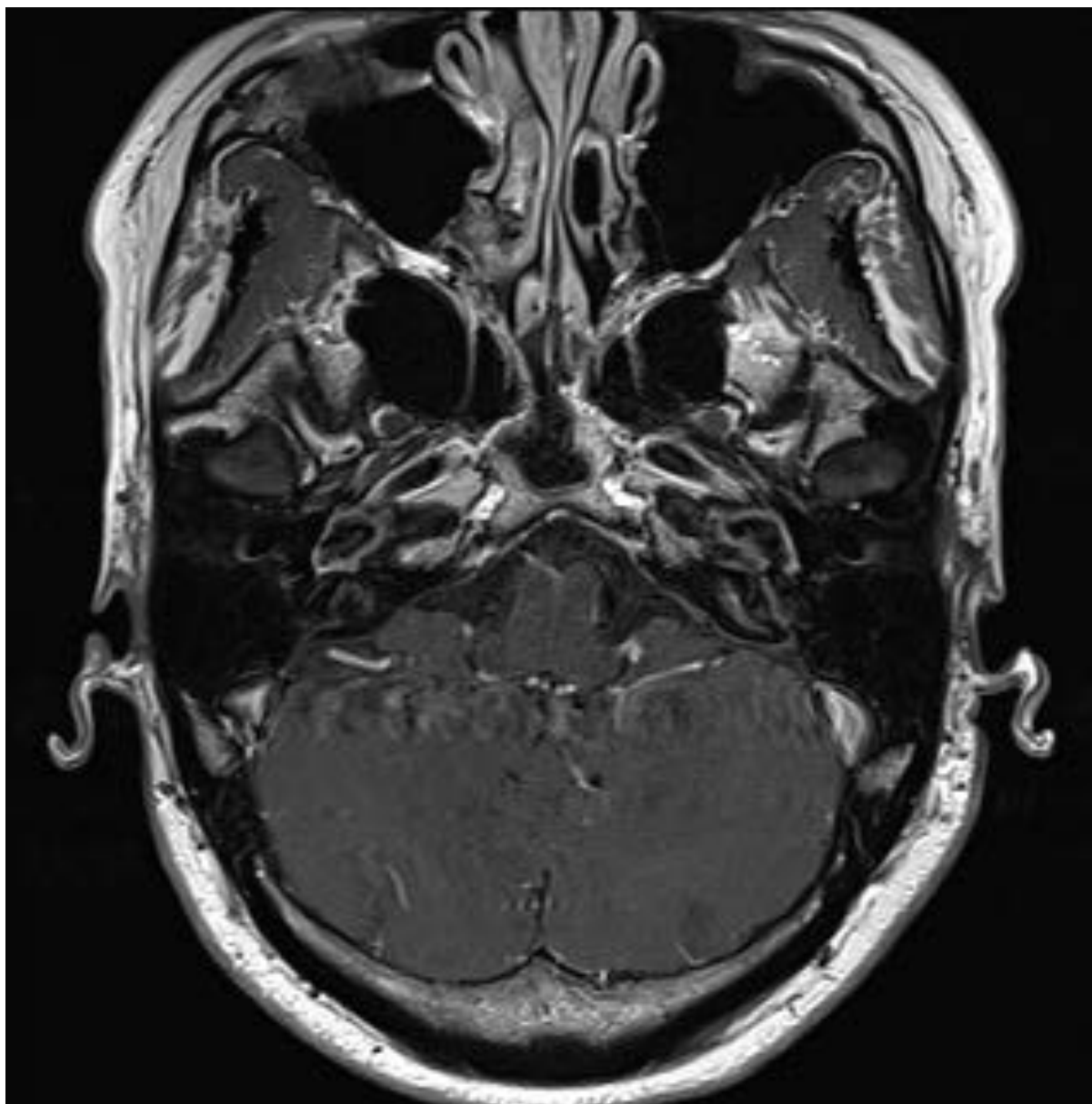












Other bone remodeling diseases

- Osteogenesis imperfecta^{1,2}
 - Genetic disease with defects in the collagen I
 - May be indistinguishable from otosclerosis
 - Can lead to sensorineural, mixed or conductive HL
- Fibrous dysplasia²
- Osteopetrosis²
- Osteitis deformans (Paget's disease)²

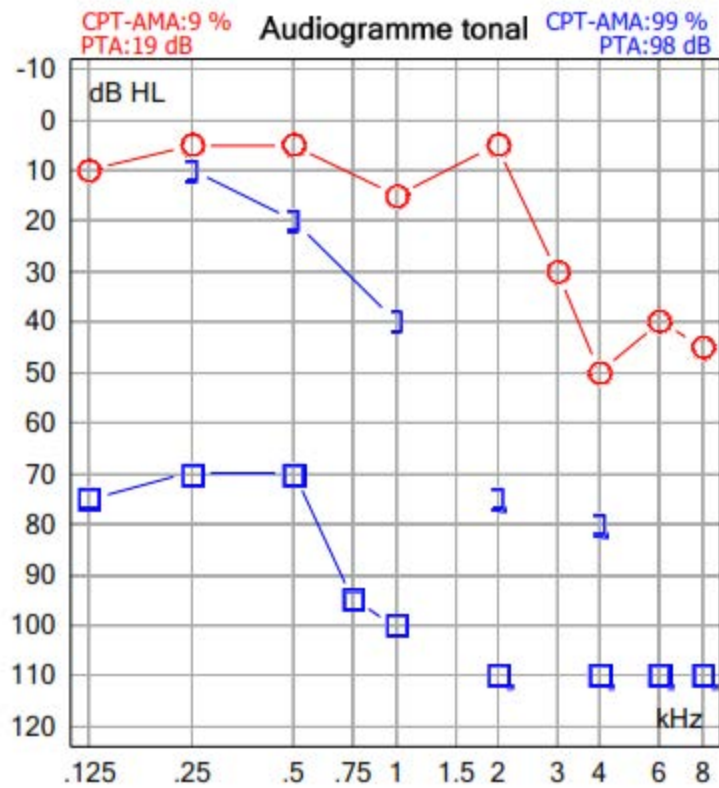
¹Santos F1, McCall AA, Chien W, Merchant S. Otopathology in Osteogenesis Imperfecta. Otol Neurotol. 2012 Dec;33(9):1562-6

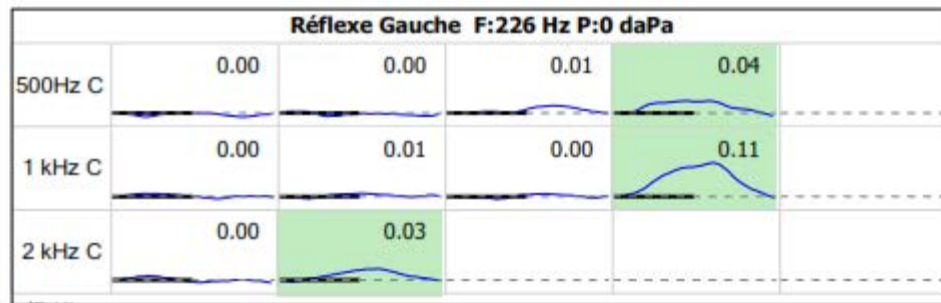
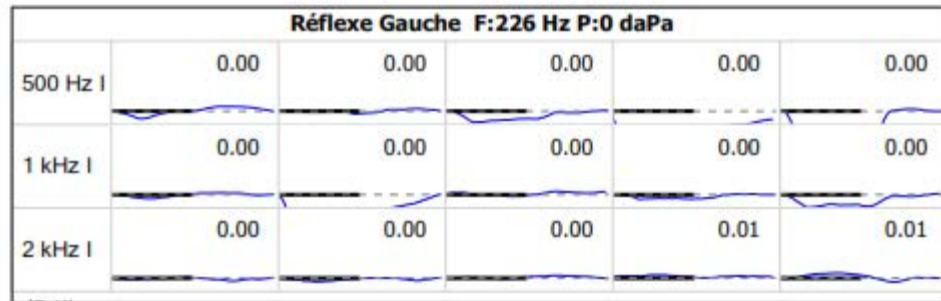
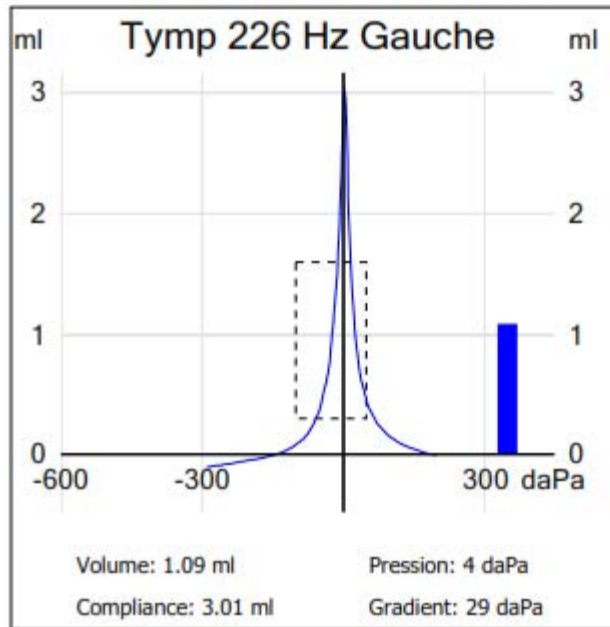
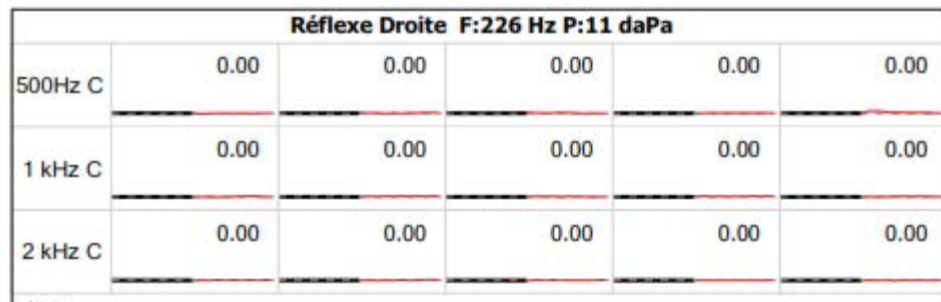
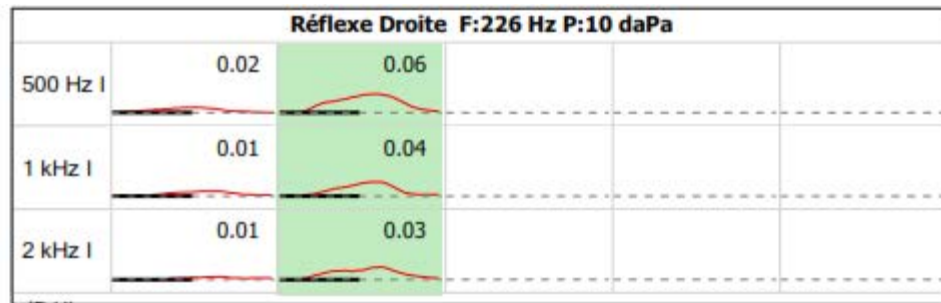
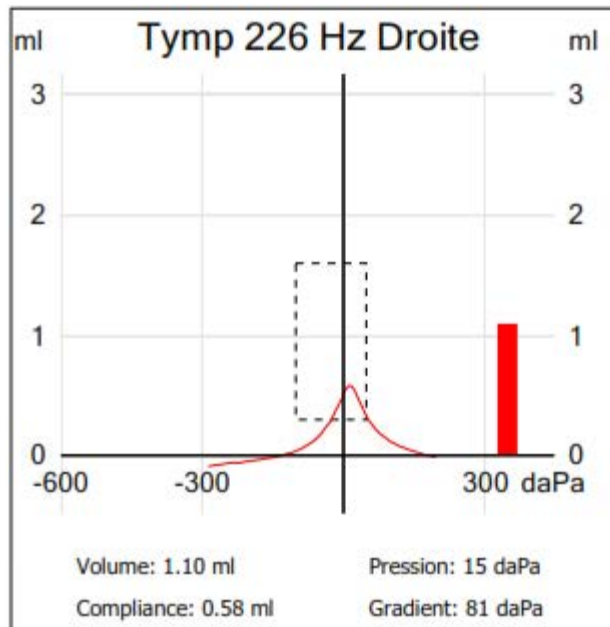
²Schuknecht's Pathology of the Ear, 3rd edition, 2010

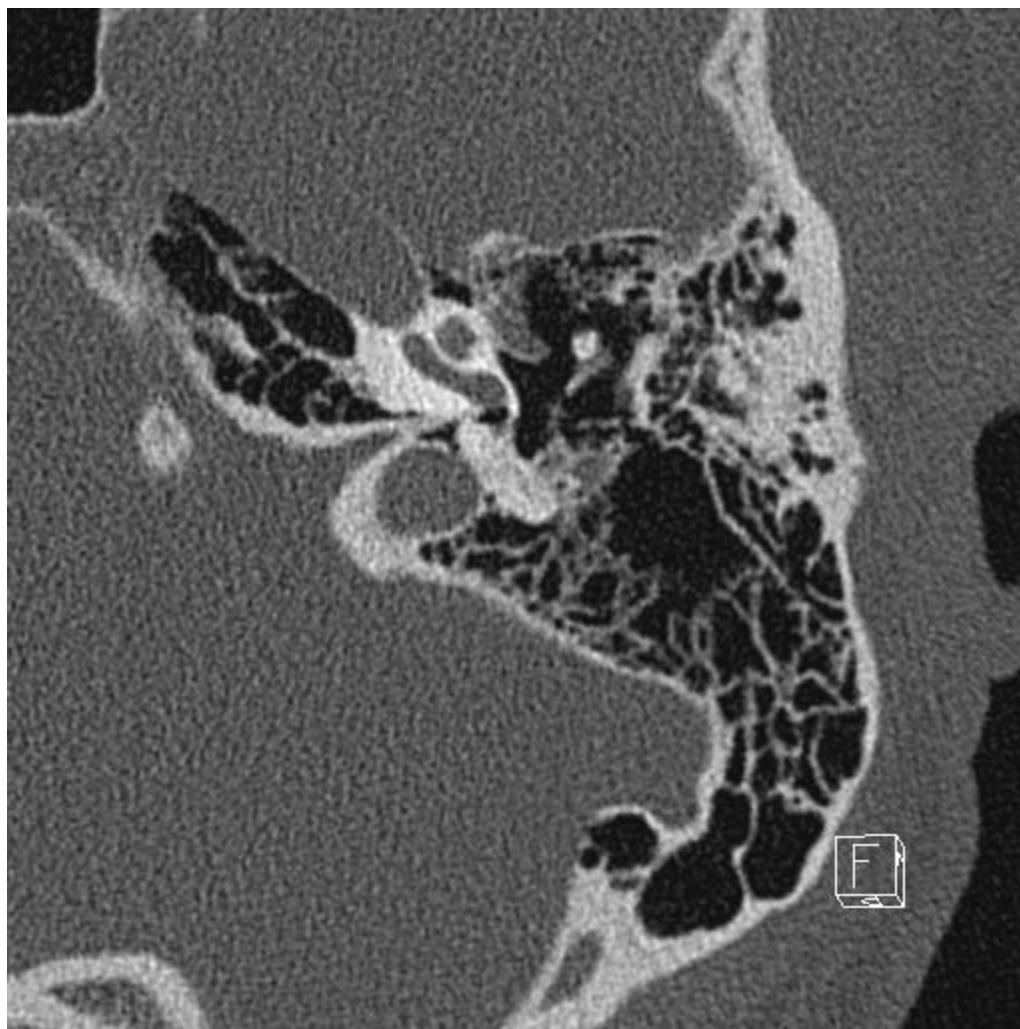
Differential diagnosis of conductive and mixed hearing loss: diseases without bone remodeling of the temporal bone

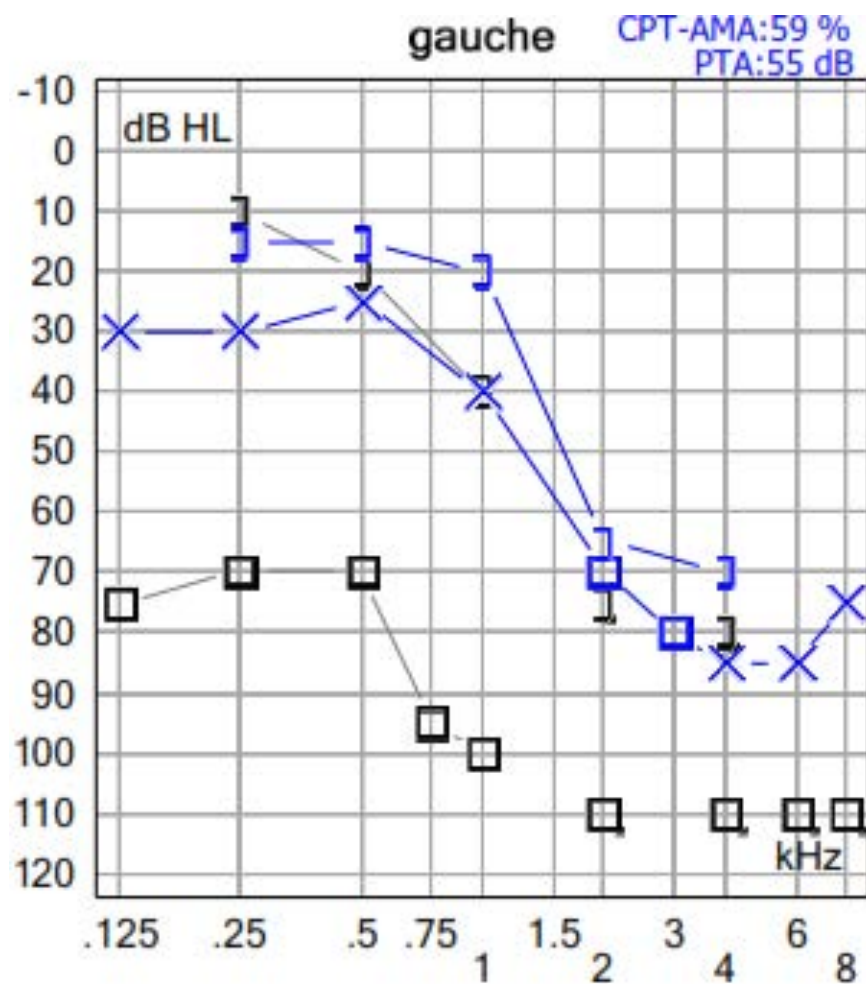
57-year old physician

- Bobsled accident as a child and since then no functional hearing on the left side
- Small sudden sensorineural hearing loss on the right side 3 months ago, anxious to lose hearing also on the right side
- Weber to the right, Rinne positive on the right and negative on the left. Membrane intact on both sides.









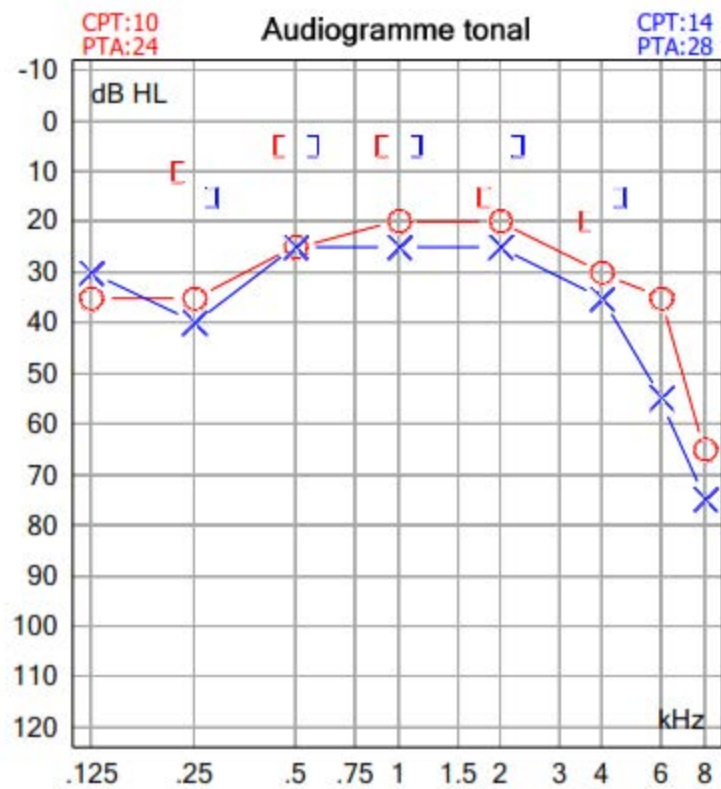
Trauma of the temporal bone

- Temporal bone fractures (longitudinal, transverse and mixed) lead to sensorineural, conductive and mixed hearing losses
- Ossicular injuries may occur with or without temporal bone fractures
 - The most common injury is the subluxation of the incus with separation of the incudo-stapedial joint
 - Therapeutic options
 - hearing aid
 - ossicular reconstruction
 - combination ossicular reconstruction and hearing aid
 - cochlear implants

¹Schuknecht's Pathology of the Ear, 3rd edition, 2010

58-year old construction worker

- Working accident with fall, brain commotion, no temporal bone fracture
- Diminished hearing, echo hearing, perceives body sounds too loud (footsteps)
- Imbalance
- Valsalva-induced vertigo

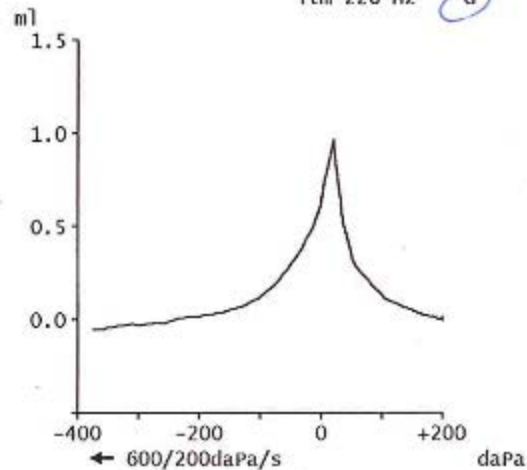


DIAGNOSTIC TYMP

TEST 1

Ytm 226 Hz

G



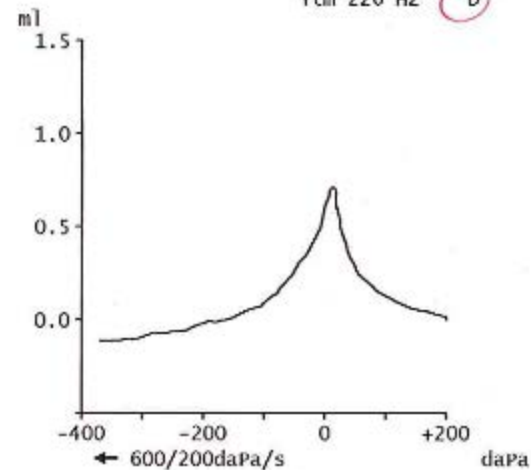
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 TYMP 1:
 TYMP 2:
 TYMP 3:

DIAGNOSTIC TYMP

TEST 4

Ytm 226 Hz

D

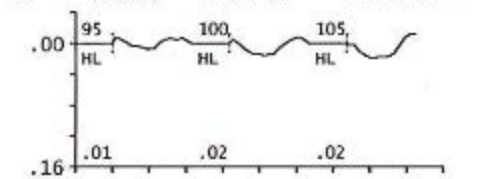


VOL COND AUDIT: 1.3 daPa ml 0.7
 TYMP 1:
 TYMP 2:
 TYMP 3:

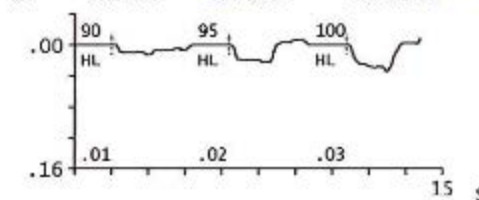
SEUIL REFLEXE

TEST 2

Y 226Hz ON:1.5 I 1000 Hz daPa:20 G
 OFF:1.5



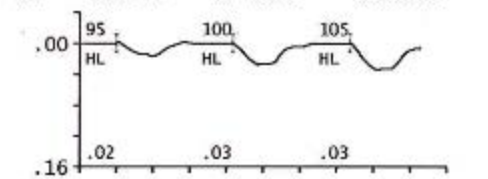
Y 226Hz ON:1.5 CS 1000 Hz daPa:20 G
 OFF:1.5



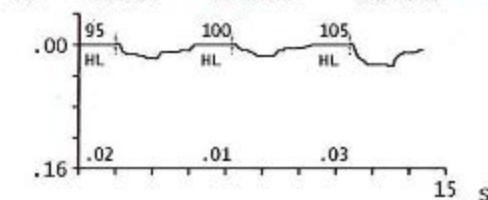
SEUIL REFLEXE

TEST 5

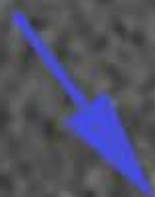
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 OFF:1.5

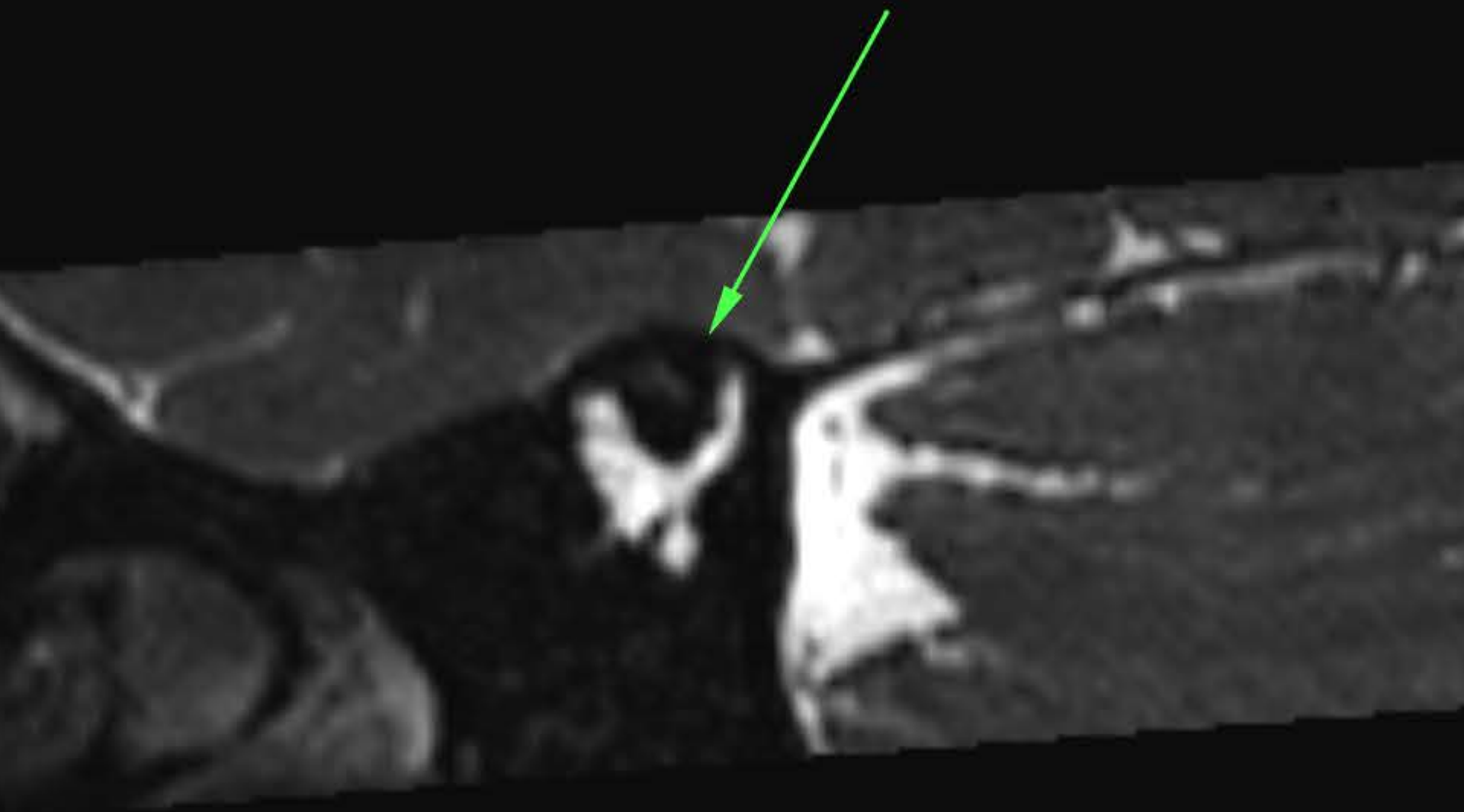


Y 226Hz ON:1.5 CS 1000 Hz daPa:15 D
 OFF:1.5



dehiscence CSS gauche





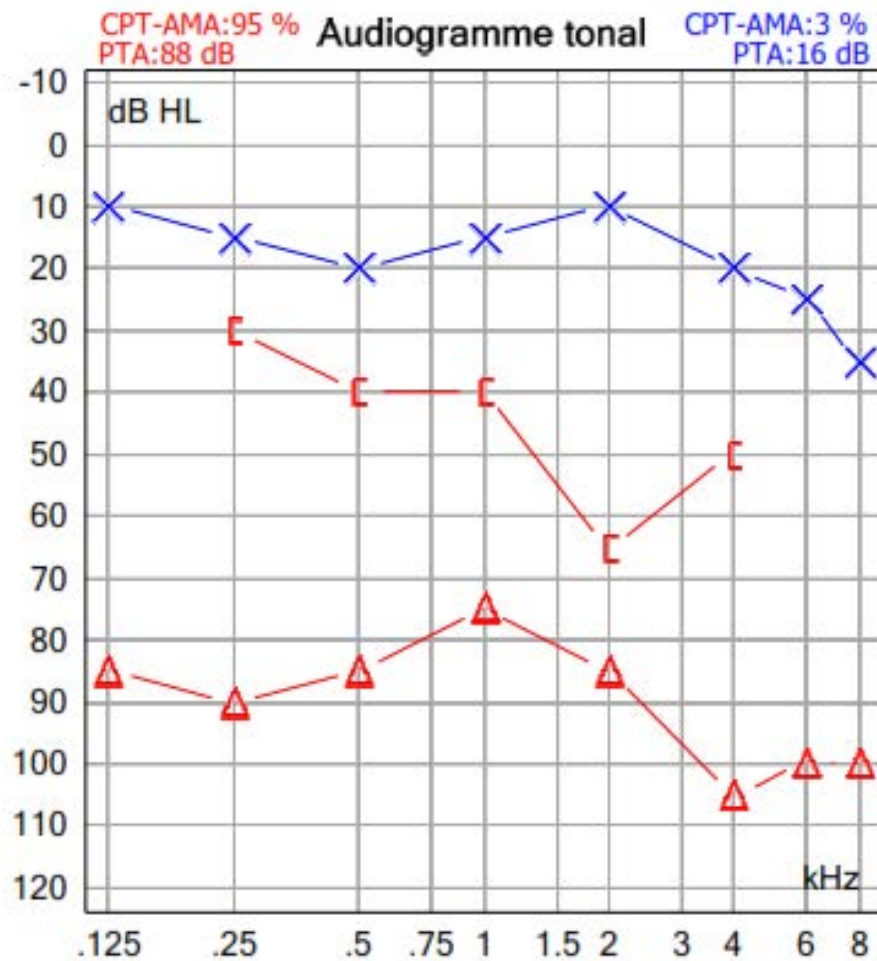
Superior semicircular canal dehiscence syndrome

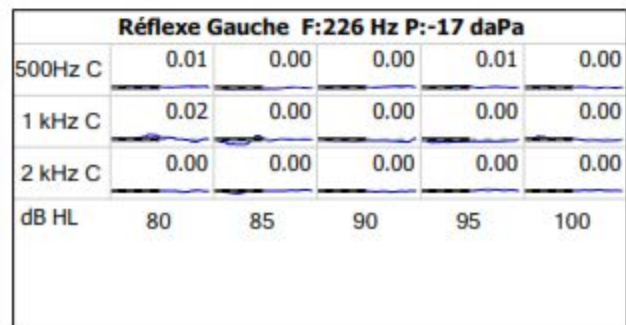
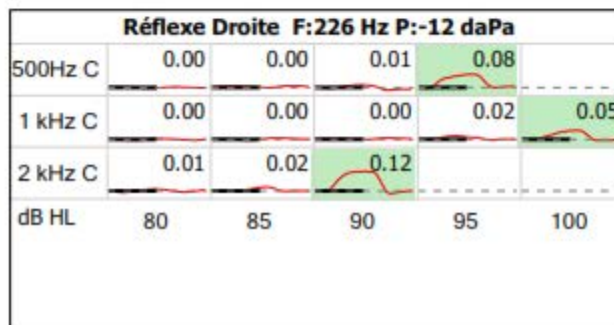
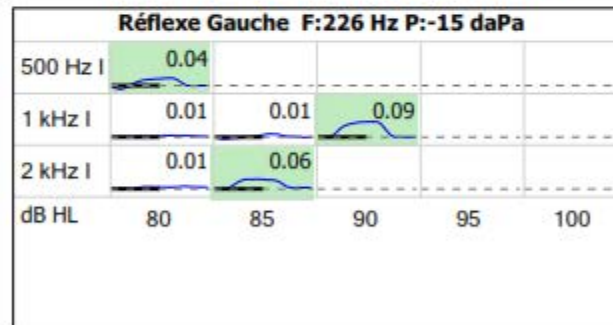
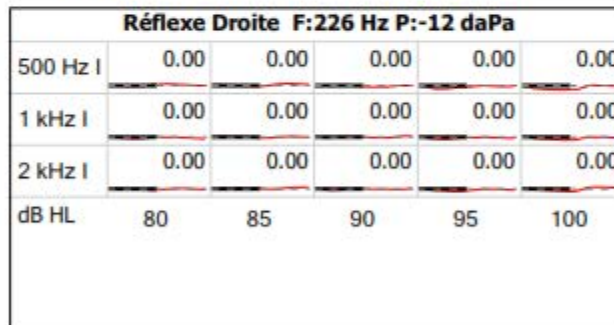
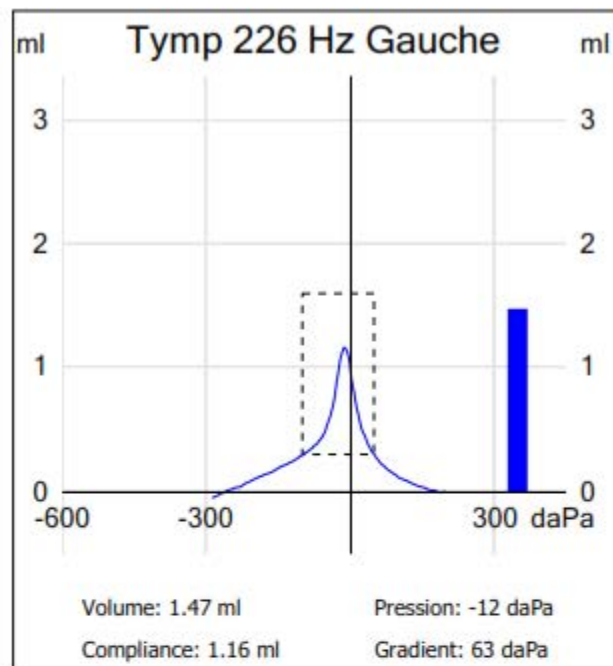
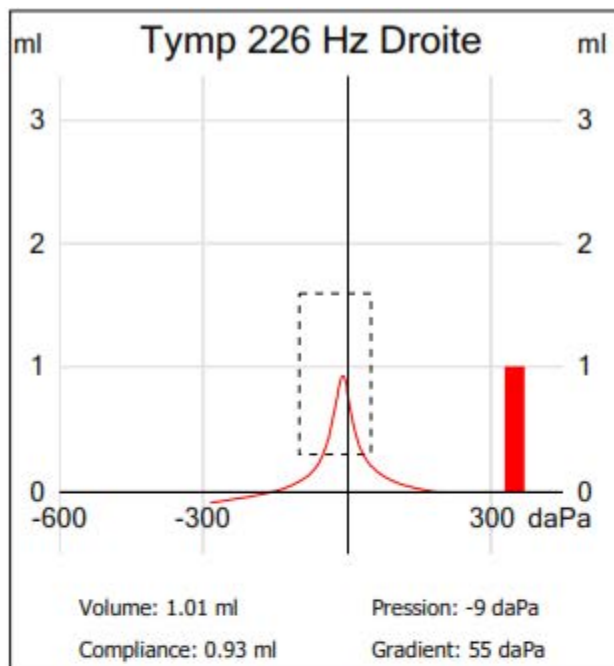
- Third mobile window of the otic capsule leading to loss of acoustic energy from the inner ear¹
 - Hearing loss and intolerance to certain sounds
 - Noise and pressure-change induced vertigo (with oscillopsia in some patients)
 - Air-bone gap without ossicular fixation (stapes reflex present)
 - Diagnosis of ssc dehiscence made with CT or MRI
 - Ssc dehiscence syndrome = clinics + radiology

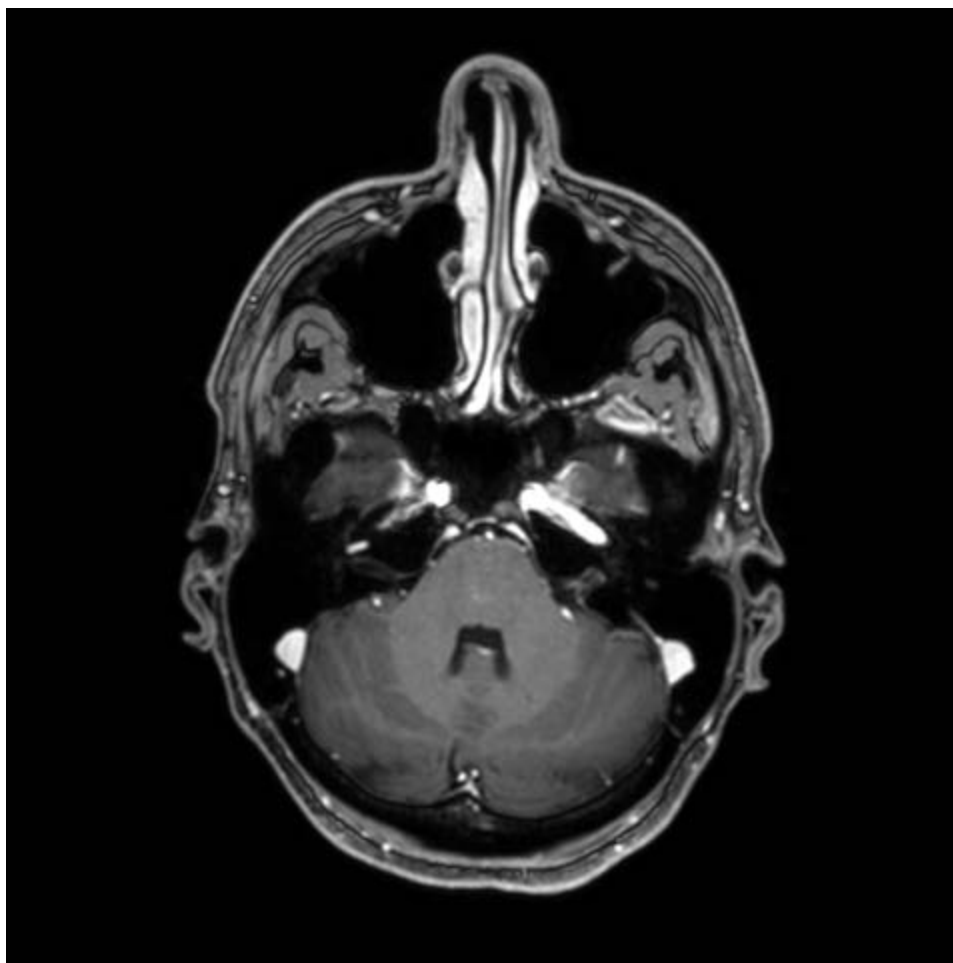
¹Minor LB et al. Ann N Y Acad Sci. 2001 Oct;942:259-73. Symptoms and signs in superior canal dehiscence syndrome.

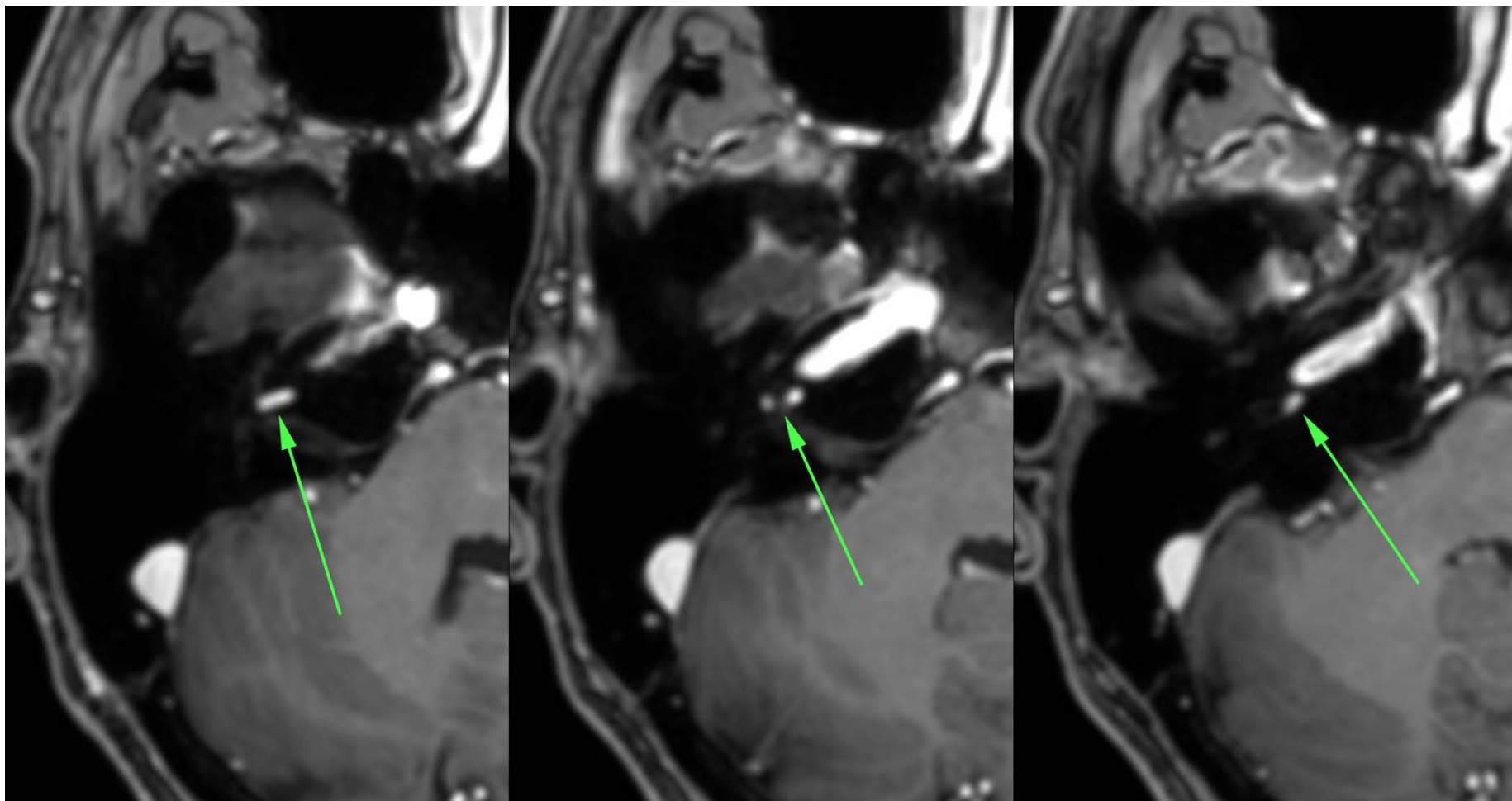
28-year old handicaped epileptic

- Mother recognizes that he stops using the right ear for telephone use from one day to another
- No complaints from his side (does not express himself without being asked)
- Status: Weber to the left, Rinne negative on the right, positive on the left, tympanic membrane intact, non-irritated on both sides









Intralabyrinthine Schwannoma¹

- Rare, benign tumor arising from the peripheral branches of the vestibular or cochlear nerve
- Typically leads to sensorineural hearing loss and occasionally to mixed hearing loss (as in the presented case)
- Therapeutic options
 - Wait and scan
 - Radiosurgery
 - Surgical resection

¹Intralabyrinthine Schwannomas. Otol Neurotol 2003; 24:299-307

Summary

- Non-inflammatory pathologies of the middle (and inner) ear may lead to hearing loss
 - Otosclerosis (frequent – 1% of whites affected)
 - Other bone remodeling diseases (rare)
 - Trauma
- Inner ear pathologies mimicking ossicular pathologies (rare):
 - Semicircular canal dehiscence syndrome
 - Intralabyrinthine Schwannomas

Conclusion

- The anamnesis of hearing loss without ear infections and the status with an intact tympanic membrane lay the base for a straight forward short list of possible non-inflammatory middle (inner) ear pathologies
- Audiometric evaluation including **stapedial reflex measurements** are crucial
- Temporal bone imaging very useful for diagnosis and for planning of treatment